BRAIN.

PART 4, VOL. 44.

DISTORTIONS OF THE VISUAL FIELDS IN CASES OF BRAIN TUMOUR.

(Sixth Paper.)

THE FIELD DEFECTS PRODUCED BY TEMPORAL LOBE LESIONS.[1]

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INTRODUCTION.

TEN years ago when this series of papers[2] was projected, accurate perimetry as we understand it to-day was rarely employed; and, indeed, as an instrument of precision the perimeter was rather an ornament and object of curiosity than an instrument of daily use in ophthalmological and neurological clinics.

To be sure, for the sake of record, the fields were occasionally plotted when the presence of a hemianopsia was more or less obvious,

[1] Presented before the American Neurological Association, Atlantic City, June 14, 1921.

[2] The preceding numbers have been as follows:—
First Paper (with George J. Heuer): "Statistical Studies." *Johns Hopkins Hosp. Bull.*, 1911, **22**, 190-5.
Second Paper (with George J. Heuer): "Dyschromatopsia in Relation to Stages of Choked Disc." *Journ. Amer. Med. Assoc.*, 1911, **57**, 200-8.
Third Paper (with Clifford B. Walker): "Binasal Hemianopsia." *Arch. Ophthal.*, 1912, **41**, 559-98.
Fourth Paper (with Clifford B. Walker): "Chiasmal Lesions with especial Reference to Bitemporal Hemianopsia." *Brain*, 1915, **37**, 524-42.
Unnumbered Paper (with Clifford B. Walker): "Studies of Optic Nerve Atrophy in Association with Chiasmal Lesions." *Arch. Ophthal.*, 1916, **45**, 407-37.
Fifth Paper (with Clifford B. Walker): "Chiasmal Lesions with especial Reference to Homonymous Hemianopsia with Hypophyseal Tumour." *Arch. Ophthal.*, 1918, **47**, 119-45.

or when vision was impaired by a large scotoma. But the cases were infrequent and few clinicians cared to perfect themselves in the use of the instrument which was time-consuming and at best gave information of only secondary diagnostic importance.

There were, perhaps, other reasons to account for its neglect. Lesions affecting the visual pathway, whether from tumour, trauma or vascular disease, were more or less hopeless from a therapeutic standpoint, and, what is more, in the presence of tumour such field defects as might be of localizing value were usually obscured by the contractions due to the secondary optic atrophy usually present before the diagnosis of tumour was ventured upon. Then, too, pituitary tumours in which successive perimetric records are especially important were regarded as rare, and usually passed unrecognized.

All this has greatly changed, more so, I fear, than is generally appreciated, if one may judge by the imperfect readings, taken with a single test object and recorded on small charts, such as are commonly sent to us when patients are referred to the clinic. Our own early records were of this sort, but when compared with readings such as those taken by Dr. Walker while attached to the clinic, the difference is as great as that between a child's sketch and an architect's drawing to scale. One gives an impression often misleading; the other the exact details. And many of the early errors into which we were trapped, such as our mistaken views on colour-interlacing, have thereby come to be explained and avoided [6]. Dr. Walker's chief contributions to the subject, as set down in his independent papers [7, 8], lie in the direction of accuracy as well as of simplification, and of late we have come to rely for clinical purposes on quantitative perimetry with test objects graded down to the smallest visible size. By so doing we may safely disregard the recording of the colour fields, provided those for form are sufficiently exact, for, as he has stated, "Colour defects are practically always represented by form defects when the visual angle is made small enough." Though few can hope to have the experience and acquire the facility in perimetry possessed by Dr. Walker, nevertheless the time necessary for the examination may be kept within reasonable bounds and trustworthy fields of vision secured after some practice by any intelligent observer copying his methods.[1]

[1] It may be said that the perimetric charts introduced by Dr. Walker are large sheets the same size as the hospital history sheets. They have necessarily been reduced to such an extent for purposes of this publication that many of the figures indicating the size of the discs, which have been employed in plotting the fields, are illegible in the reproductions accompanying the text.—Ed.

The Temporal Loop of the Optic Radiation.

Before describing the disposition of the geniculo-calcarine fibres in the lobe, which is the matter of primary importance, the story given in the first paper in this series, as an illustration of the value of accuracy in perimetry, may deserve re-telling.

There entered the Johns Hopkins Hospital in 1910 a man subject to convulsive seizures, with a gustatory aura typical of fits of uncinate gyrus origin. Some years before he had been shot in the left eye, and, as a radiogram showed, the ball had lodged in the petrous portion of the left temporal

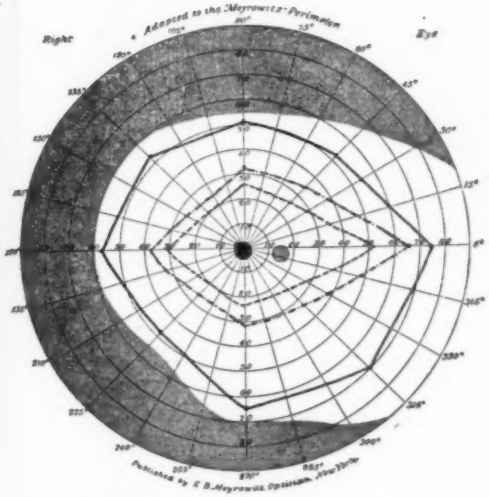


Fig. 1.—Chart of October 18, 1910, showing what were regarded as practically normal field relations. Note that eight points were relied upon for the form field and only four for the blue and red fields.

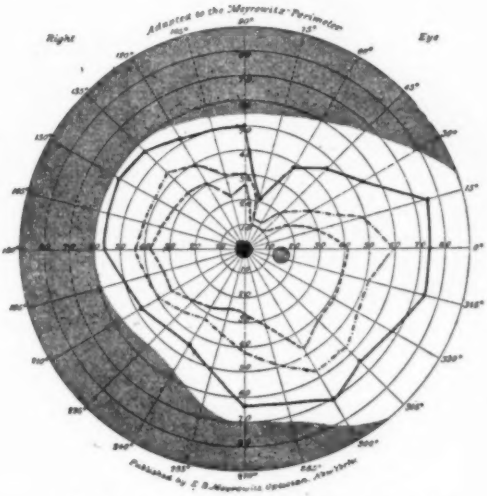


Fig. 2.—Corrected chart of October 19, 1910, for comparison with fig. 1, disclosing upper right temporal defect.

bone. Obviously the lower part of the left temporal lobe had been traversed. The field of vision of the remaining eye (fig. 1) had been plotted by Dr. S. J. Crowe, my then assistant, and it was regarded as normal. Indeed, no defect was anticipated, and the observation was made merely as the routine procedure of a painstaking house officer. The patient was shown one day on a ward visit which Dr. Adolf Meyer happened to attend, and he pointed out that the field periphery which had been plotted from tests at every thirty degrees might show a sector-shaped defect if taken with closer angulation, as only a few fibres of the pathway might have been damaged. This was done, with the result shown in fig. 2. Had the left eye not been destroyed, the defective sector would presumably have appeared as a homonymous one.

Three years before, in a paper which has received an undeserved lack of attention, Meyer [5] had described "the peculiar detour of the ventral portion of the geniculo-calcarine path which simulates the existence of an inferior longitudinal fasciculus." By the study of secondary degenerations following old vascular or traumatic lesions which had shown a fortunate limitation, either by exclusion or inclusion, to the visual pathway, he had observed that a portion of the optic radiation, on leaving the geniculate body, plunges far forward into the temporal lobe to sweep around the horn of the ventricle before it turns backward to end in the calcarine cortex. Though Meyer's findings were merely corroborative, in large part, of those by his predecessors—Flechsig, Henschen, Archambault [1], and others—he was able to demonstrate the course of isolated paths which show not only that the dorso-lateral and ventral bundles remain the same size throughout, but that they maintain in their course a definite position in relation to other bundles. Thus, the dorso-lateral bundle not only remains the most dorsal, but the most direct, all the way from the geniculate body to its end stations. The most ventral bundles, on the other hand, are the ones which make the longest detour around the temporal horn to end in the anterior part of the calcarine cortex.

A glass-model reconstruction of these fibres was made by Dr. Meyer, and it is from this that, at my request, he has permitted Max Brödel to make the superb diagrams (figs. 3, 4, and 5), which accompany this paper. They render superfluous any further written description.

Meyer's own observations were not supported or confirmed by perimetric tests, which were difficult or impossible in the clinical material of the type he had at his command. Nevertheless, he shrewdly surmised in connection with the few reported cases of quadrant hemianopsia in which small defects had been found post mortem that the dorsal bundles have an upper retinal distribution and hence correspond with field defects in the lower quadrants, whereas *per contra* involvement of the ventral bundles is responsible for defects of the upper field quadrants such as was shown in fig. 2. In other words, the disposition of the fibres in the geniculo-calcarine radiation is practically the same as that in the optic nerve itself. Whether or not this means, as I believe it does, separate representation on the cortex for different areas of the retina, need not now detain us, for our present concern lies with the course of the fibres through the temporal lobe and not with their calcarine end-stations.

From the series of cases to be reported, we shall see that tumours

FIG. 3.—Showing the brain, as transparent, to indicate the lateral ventricle and the disposition of the fibres of the optic radiation.

FIG. 4.—Mesial aspect of the brain with disposition of the geniculo-calcarine pathway.

in the temporal lobe may produce homonymous field defects in the upper or lower quadrants which are of extreme value in localization. Lesions more posteriorly situated are in my experience less likely to

FIG. 5. —The radiation shown on the right in section and on the left in transparency.

produce these partial defects, or at least are less likely to be caught by the observer in a stage short of the usual midline hemianopsia.

SOURCE OF MATERIAL.

In the writer's series of 663 verified intracranial tumours, to April 1, 1921, there have been 197 tumours involving the hind-brain

(subtentorial), 190 pituitary tumours (including suprasellar lesions) and 276 tumours involving the cerebrum proper. Of this group of 276 cerebral tumours, in 59 the lesion has been situated largely within or has chiefly deformed the temporo-sphenoidal lobe. This by no means represents all the temporal lesions, but the doubtful cases have been excluded, since for our present purposes it is best to limit the discussion to those tumours which appear to have originated in the lobe itself.

Even so, that 59 out of 276, or over one-fifth of all verified cerebral tumours, should have primarily involved the temporal lobe may appear an unduly large number. For this, two explanations may be offered. One lies in the fact that, owing to the common practice of performing a right subtemporal decompression in tumours of doubtful situation, an unexpected temporo-sphenoidal lesion has not infrequently been disclosed.[1]

Another explanation is that tumour localization in this relatively silent area has of late years become more and more exact owing to the interpretation of field defects, which this communication, it is hoped, will serve to point out.

In 39 of these 59 temporal lobe cases perimetric charts sufficiently trustworthy for our present purposes were obtained and in many instances repeated observations over long periods have been made. In the remaining 20 cases the tests were untrustworthy or impossible for a variety of reasons, owing to practical blindness from secondary optic atrophy (six cases); owing to such a degree of stupor or mental confusion as to preclude dependable responses (eight cases); owing to other causes as aphasia, childhood, &c. (six cases). The fields of vision in the 39 dependable cases may be subdivided as follows into those showing:—

[1] It may need emphasizing that the cranial defect in a subtemporal decompression is made below the attachment of the muscle to the temporal ridge and consequently it is unusual to have more than temporal convolutions exposed. Occasionally one may barely expose the Sylvian vessels, but when there is a tumour of the lobe the Sylvian fissure is apt to be dislocated upward.

It is significant (1) that in the fifty-nine cases the lesion was found on the right side in thirty-two and on the left in twenty-seven instances; (2) that in only sixteen of the thirty-two right-sided cases was the lateralization helped out by perimetry. Some of the remaining sixteen had uncinate attacks and contralateral facial weakness assuring the localization, but the others were stumbled upon in the course of a routine decompression.

In a correspondingly large series of unverified tumours there is a long list of undoubted temporal lobe cases which have been excluded from this discussion. Many of these cases had uncinate seizures and field defects comparable to those in the verified cases, to the discussion of which this paper will be limited.

(1) *No defect* (*six cases*).—In these the fields were at all times normal. Three of them were slow-growing endotheliomata and one an angioma deforming the lobe from without, and therefore less likely to affect the pathway. That field defects may occur under corresponding conditions, however, some of the cases to be reported later on will make clear. A fifth case was a rare one of dermoid cyst in the tip of the lobe accompanied by uncinate seizures. From the situation of the lesion one would have expected a partial hemianopsia which repeated examinations failed to reveal. The sixth case was a glioma disclosed by a right decompression in which only a single preoperative test had been made.

(2) *Homonymous hemianopsia* (*eight cases*).—In these the condition had advanced to a median vertical separation of blind and seeing fields, indicating an interruption of the entire pathway, though the macular fibres were oftentimes spared. In none of these cases had fields been plotted sufficiently early to detect quadrantic defects, nor in any of them was a post-operative recession from the hemianopsia observed. The findings indicated that the cerebral lesion was on the right in five and on the left in three of these cases, but it did not otherwise indicate the position of the tumour in the hemisphere. It is presumable that in many or all of them the growth may have come to involve the geniculate body or even to have affected the optic tract itself by pressure. If reliance is to be placed on the sparing or otherwise of the macular bundle as an indication of the involvement of optic radiation *versus* optic tract it would appear that tract involvement may be expected as a late feature of all temporal lobe tumours. In four of these eight cases the macula was included in the field bisection at the time of the examination and in all likelihood in the course of time the macula came to be involved in all. Several examples of hemianopsia, both with macula spared as well as included, which occurred in the series, have been classified under the following heading for the reason that the patients showed at one time or another the partial defects (e.g., Case 7) with which we are chiefly concerned.

(3) *Partial hemianopsia* (*twenty-five cases*).—It is upon these defects, more or less quadrantic in character, that chief emphasis will be laid. At one time or another while under observation the patients all showed an homonymous defect, which fell short of a complete hemianopsia. As will be indicated by the case histories which have been selected for purposes of illustration, these partial defects have been observed either as: (1) a stationary condition; (2) a con-

dition advancing toward a hemianopsia; or (3) a condition in process of recession after a successful operation.[1]

Case Reports.

At the outset one or two examples of some of the earlier cases in the series may be given. The fields were then taken with standard 5-mm. test objects, and many of the details of the more recent examinations are therefore wanting. These early observations, nevertheless, served to give us our first hints of the localizing value of these partial defects. This was particularly true of defects in the colour fields which were seen to precede those for form, a matter which is of less importance to-day, since practically the same information is secured by utilizing white discs of minute size. It was from some of these early cases, furthermore, that we first came to appreciate the fact that the field defect in the eye on the side of the lesion is apt to be in advance of the other.

Case 1.—*Gliomatous cyst of right temporal lobe producing a lower homonymous defect short of hemianopsia. Post-operative widening of fields to normal.* (J. H. H., Surg. No. 23551.)

January 30, 1909: Admission of S. G. W., a clerk, aged 26, with the complaint of headaches, fainting attacks and failing vision.

Chronology of symptoms: Always well until July, 1905, when he had a sudden loss of consciousness without warning or convulsive movements. One year later another similar attack, preceded by dizziness. Since then he has been "nervous" and somewhat less alert intellectually.

[1] On two earlier occasions, in 1912 with Dr. Heuer, and again in 1916 with Dr. Walker, preliminary steps were taken to assemble material for this paper, and to review the literature pertaining thereto. The first interruption came from my transfer to Boston, and the second was due to our entry into the war. To have been deprived of their collaboration is by no means made up for by the fact that the number of cases which can be drawn upon has greatly increased. My acknowledgments are due not only to them for a great deal of the preliminary work, but also to a succession of neuro-surgical assistants who will recognize many of their own charts and case reports in the text.

Our studies in 1912 were chiefly directed toward a determination of the value of Wernicke's hemiopic pupillary reaction, and also of Wilbrand's prism test in differentiating, in cases of pure hemianopsia, between an interruption of the pathway central or peripheral to the geniculate body. The tests, though sharply positive in some cases, were so obscure in others, that we subsequently have abandoned them, for at best the information they give is but confirmatory of what one may gain in other ways. Moreover, it is highly desirable to recognize and interpret field defects before a total hemianopsia has supervened and at that time these tests are not applicable. A homonymous hemianopsia, whether or not the macula is spared, which is accompanied by a primary optic atrophy, one may safely ascribe to a chiasmal lesion usually pituitary in origin: one which is accompanied by a choked disc probably involves the radiation somewhere in its course. This is not an invariable rule, but it is less likely to mislead than a dependence on the Wernicke and Wilbrand tests.

In August, 1908, onset of persistent headaches, chiefly nocturnal, increasing in severity. At about this time some disturbance of vision was first noticed, with occasional diplopia. Also, from time to time, flashes of light described as "luminous things" have been seen, but they are not lateralized. Also vague "dizzy spells" without loss of consciousness, but with no features suggesting an uncinate origin.

Physical examination: This was quite negative except for: (1) a choked disc of 3 D. right, with beginning atrophy and $\frac{20}{60}$ vision: 4 D. left, with $\frac{20}{30}$ vision; (2) an incomplete left homonymous hemianopsia (fig. 6). No hemiopic pupillary reaction (Wernicke).

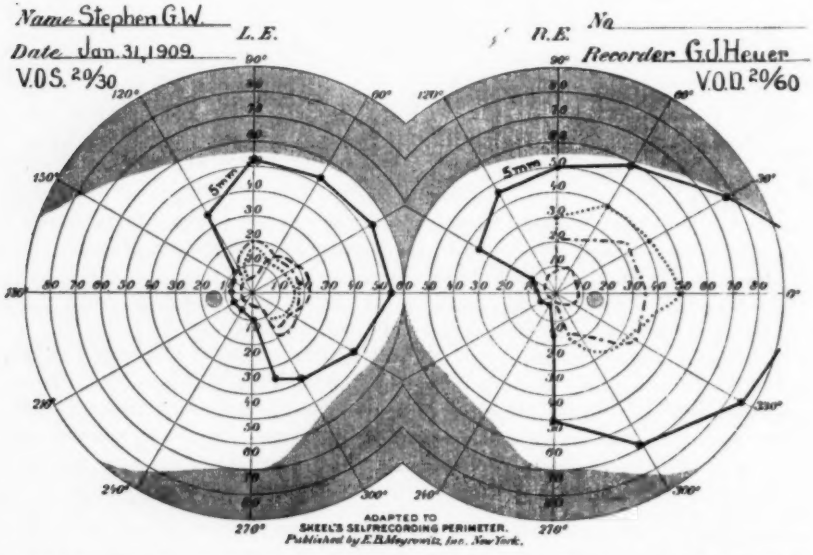


FIG. 6.—*Case* 1. Fields before operation which disclosed a gliomatous cyst of the right temporal lobe.

February 2, 1909: Operation.—Right subtemporal decompression. This disclosed a dry bulging temporal lobe which from surface appearances contained a subjacent lesion. A large gliomatous cyst pointing in the upper temporal convolution was widely opened and was drained for a week.

Post-operative: An immediate cessation of pressure symptoms. The choked discs subsided with gradual restoration of visual acuity to normal. Fields were taken every few days after February 10, on which date they showed normal outlines for form, though the colour fields were still slightly contracted (e.g., fig. 7). He was discharged March 4 with normal fields and vision of $\frac{20}{18}$ in each eye.

Subsequent note: Three years later (February 19, 1912) he returned for observation. He had been free from symptoms during the interval. The fields remained normal. After this the patient passed out of my hands. Two years later—nine years from the time of his primary loss of consciousness—he died with a recurrence of tumour symptoms.

Comment: When this patient was first seen, without the perimeter even a lateralizing diagnosis would have been impossible. It may be observed in the first charts (fig. 6) that there was a hemiachromatopsia on the right, the side of the lesion, the macula being spared. Doubtless the primary field defect began in the lower left quadrants, but it is improbable that the significance of this was sufficiently appreciated twelve years ago to justify us in anticipating the temporal lesion which was stumbled upon.

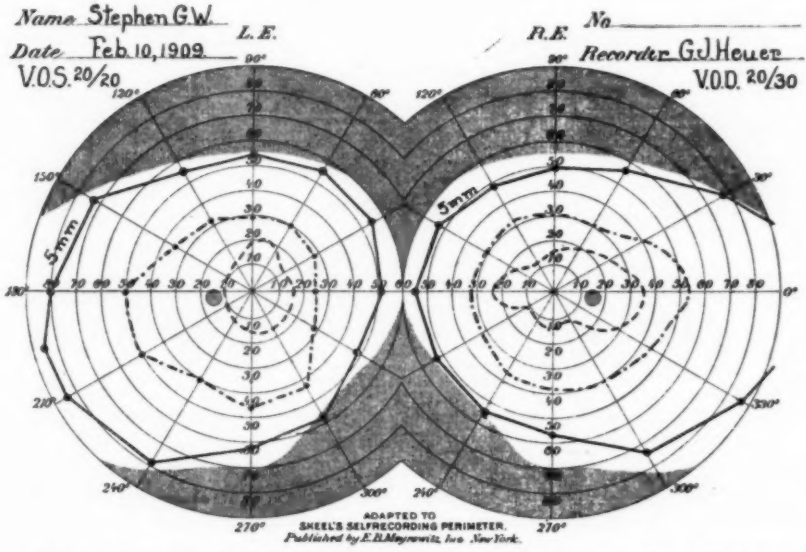


FIG. 7.—*Case* 1 (*contd.*). Fields eight days after operation, for comparison with fig. 6.

In the following case the conditions so far as the diagnosis was concerned were much the same as the above, though produced by a tumour of different type—a meningeal endothelioma. The case is one which interested us greatly at the time and was studied in detail, though the bare outline of the history must suffice for this report.

Case 2.—Large endothelioma involving right temporo-sphenoidal lobe producing a homonymous field defect unequal in the two eyes. Operation: Partial recession of field defect. (J. H. H., Surg. No. 30210.)

July 9, 1912: Admission of Louis C., a tailor, aged 33, complaining of headaches and failing vision.

Tumour history: For three years mild but more or less continuous headache referred to the right parietal region. For three months progressive failure of vision in the right eye. No other subjective disturbances. He had worked until the day of admission.

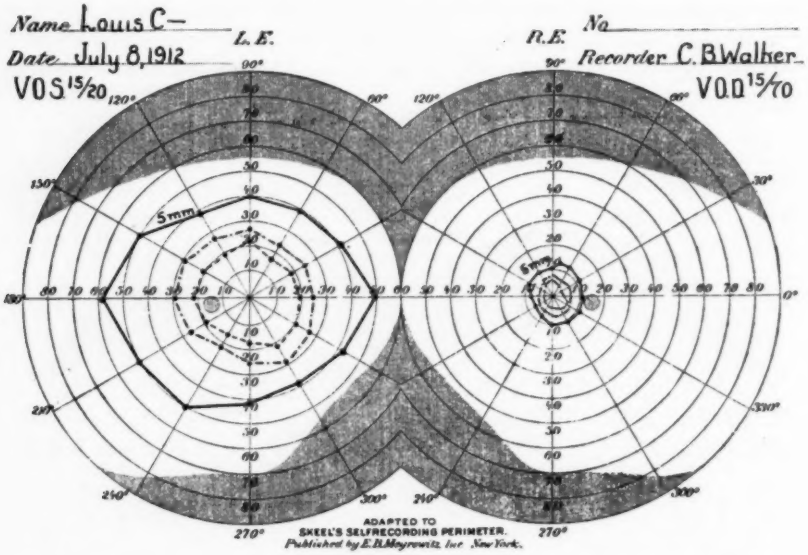


FIG. 8.—*Case* 2. Fields taken before operation which disclosed endothelioma of right temporal lobe.

Physical and neurological examination: Negative in all respects except for the condition of his eyes, which were slightly exophthalmic. There was a choked disc of 3 D. right and 2 D. left, with new tissue and some secondary atrophy, more marked right than left. The fields showed (fig. 8) concentric constriction, slight on the left but extreme on the right, where vision was practically tubular. V.O.S. $\frac{15}{20}$; V.O.D. $\frac{15}{70}$. The X-ray showed signs of general pressure but nothing of localizing value.

July 11, 1912: Operation 1: A right subtemporal decompression performed by my then assistant, W. E. Dandy, unexpectedly disclosed the edge of a tumour occupying the lower anterior part of the temporal lobe.

Post-operative: He made a good recovery with partial subsidence of his

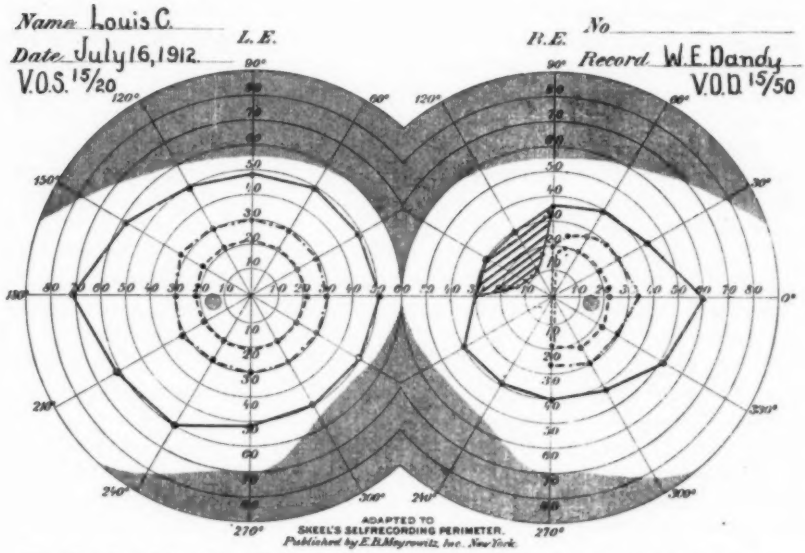


FIG. 9.—*Case* 2 (*contd.*). Fields five days after decompression which disclosed tumour. (Cf. fig. 8.)

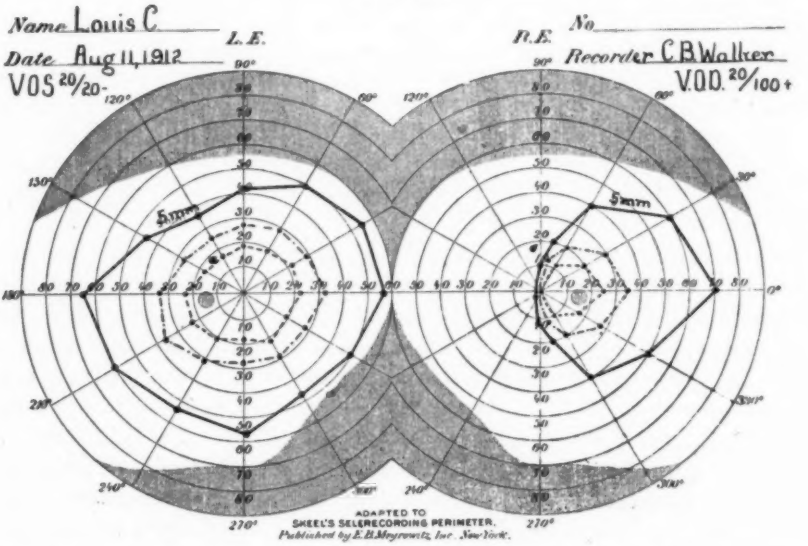


FIG. 10.—*Case* 2 (*contd.*). Fields one month after those shown in fig. 9. Tumour as yet not removed. Note beginning upper temporal defect on the left with complete right hemianopsia.

choked disc and lessening of his headaches. On July 16 the fields, which had widened, showed a nasal hemiachromatopsia with an upper form defect in the homolateral eye (fig. 9). He refused further operation and was discharged on this day to his home near by, but returned frequently for purposes of record.

By July 29 there was a fully established hemianopsia for both form and colour in the right eye, where further elevation of the disc had occurred. There was thought to be both a positive Wernicke and Wilbrand pupillary reaction, but negative by the method of Hess.

By August 11 the upper temporal field in the left eye had begun to be affected (fig. 10), indicating the beginning of a homonymous defect. His visual acuity on the right had become so much impaired that he expressed a desire to re-enter the hospital for further operation. The only additional symptoms observed and recorded at this time were a slight weakness of the left face and some subjective tinnitus referred to the left ear. The decompression area was tense and the discs measured about 2 D.

August 17 and August 26, 1912: Operation 2. Two-stage enucleation of a large endothelioma deforming the lower and anterior portion of lobe. He made a perfect surgical recovery from both procedures. On August 23, between the two sessions and before the tumour removal, the fields showed a still more marked upper temporal defect for the left eye. This was present also on August 29, three days after the extirpation, though by this time the fields had begun to widen out again (fig. 11). Fortunately, with subsidence of the choked discs normal vision returned for the left eye, though on the right at the time of his discharge the nasal hemianopsia persisted.

Subsequent note: Three years later, July 15, 1915, the fields were again taken at my request by Dr. Dandy (fig. 12). They show that in the interval there had been a slight filling out of the lower nasal quadrant in the affected eye. Four years later, October 27, 1919, visual fields kindly taken for me by Dr. Lloyd B. Whitham correspond exactly with those previously taken by Dr. Dandy. The process, therefore, may be regarded as stationary.

Comment: This case resembled the foregoing in that the examination revealed merely the general pressure symptoms of tumour and nothing whatever of localizing value if we except what the perimeter disclosed. Attention may be called to a number of interesting points.

(1) I am at a loss as to what interpretation to put on the unilateral tubular vision (cf. fig. 8), a condition commonly ascribed to hysteria. Four, or possibly five, other examples of the same condition, always on the side of the lesion, have been observed in the series of temporal lobe cases, and though we once felt that the finding might have some significance, I hesitate to lay any great stress upon the matter. (2) Of greater importance, though not observed till the tumour diagnosis had been made, was the gradual onset of the field defect in the homo-

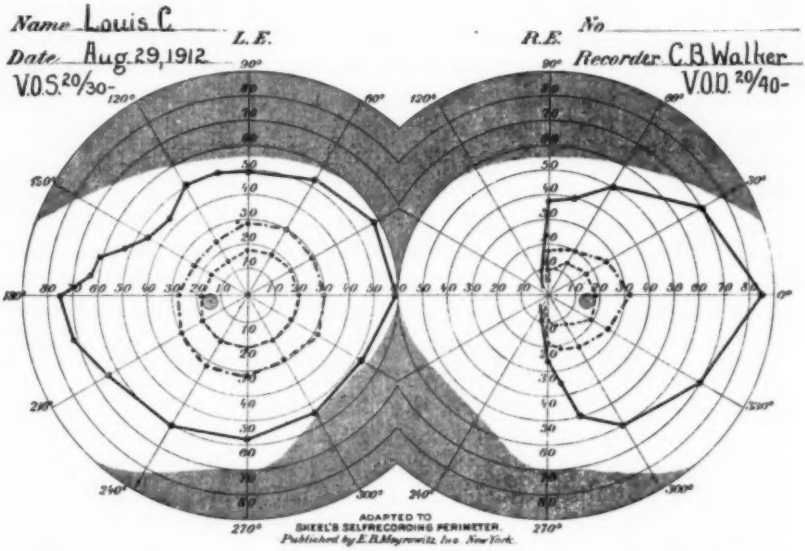


FIG. 11.—*Case* 2 (*contd.*). Three days after tumour extirpation. Note widening of fields compared with fig. 10.

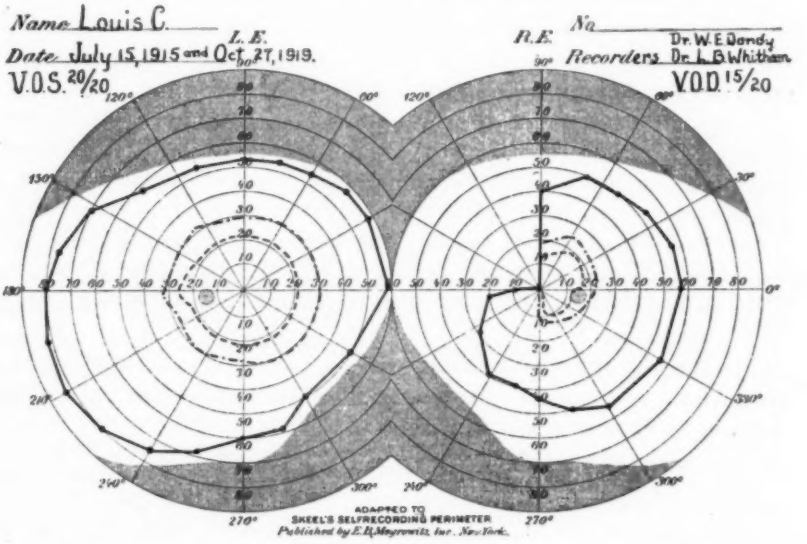


FIG. 12.—*Case* 2 (*contd.*). Present and stationary condition seven years after the tumour extirpation.

lateral eye, first showing as a hemiachromatopsia and advancing to a complete hemianopsia before there was any sign of a defect in the left eye. (3) There can be little doubt but that with graded discs these defects would have been much more apparent even in the left eye, and that a hemianopsia to form would have shown itself in correspondence with the hemiachromatopsia observed on July 16 (fig. 9), had a small test object been used below a millimetre size.

Confession must be made of one possibility of misinterpretation of these findings, particularly in consideration of the nature of the tumour and its extracerebral situation. For it is conceivable that the right optic nerve may have been pressed upon, a possibility made the more likely by the fact that the macula was impaired and there was a positive Wernicke reaction in the affected eye.

In answer to this possible criticism it may be said that pressure against the optic nerve by such a large tumour could easily have produced blindness but hardly a hemianopsia, and if the chiasma had been affected the field defects would have been more nearly equal in the two eyes. Moreover, as will be seen, a hemiopic pupillary reaction in our experience is not very dependable and the patient showed at the same time a positive reaction to the prism test which would indicate an involvement of the radiation alone. It possibly, however, is splitting hairs to discuss these differences, for, after all, the question of localization would be much the same, whichever interpretation is put upon causation of the field defects in such a case.

In the tumour cases subsequent to 1912, the more accurate perimetric methods developed by Dr. Walker have come to be utilized and have been applied to twenty-three of the thirty-nine temporal cases in which fields have been taken. In selecting the following examples from this later series, the attempt has been made to choose cases with lesions of different kinds in different situations in the lobe, as well as ones which illustrate the methods of advance as well as of recession of the field defects.

In the first of the cases which will be cited from this later series a gliomatous cyst was the offending agent. It was low-lying and, as will be seen, implicated the ventral fibres of the loop.

Case 3.—*Gliomatous cyst of right temporal lobe producing left upper homonymous quadrantopsia. Operation. Recovery.* (P. B. B. H., Surg. No. 582.)

November 14, 1913: Admission of Miss E. A. J., aged 43, with the complaint of headache and diplopia.

History: Subject to frontal headaches all her life. For six months there

have been severe attacks of pain referred to the suboccipital region. For four months considerable vertigo. Of late, some unsteadiness of gait which she ascribes to her diplopia. Also of late, paræsthesia of extremities and great despondency through a morbid fear of insanity.

On September 18, two months before her entrance, fields of vision taken by C. B. Walker disclosed a left upper quadrantal hemianopsia for form and colours (fig. 13). A month later, October 17, 1913, Dr. George S. Derby found the defect to remain practically unaltered.

Physical examination: Considerable exophthalmos, more right than left; a slightly enlarged thyroid.

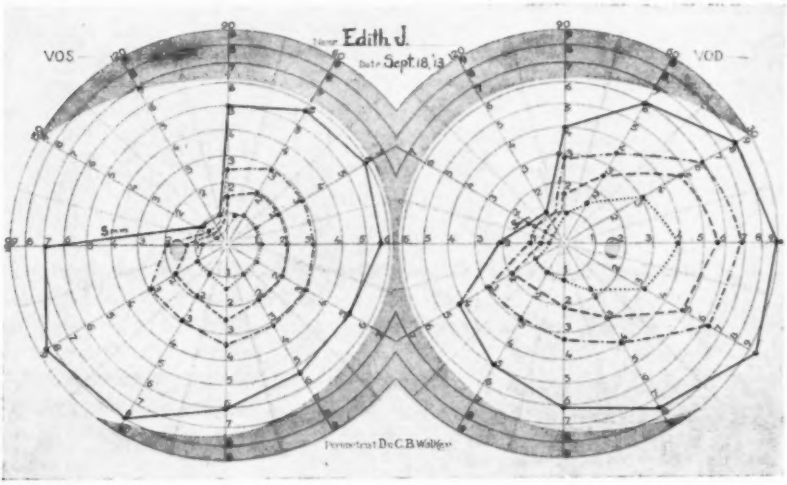


FIG. 13.—*Case* 3. Fields as found practically stationary during the two months before operation.

Positive neurological findings: A complete abducens paralysis. The eye-grounds showed slight hyperæmia with haziness of the nasal margins and overfilling of the veins; no measurable swelling. The fields taken by Gilbert Horrax showed approximately the same defect observed on the previous tests. The condition, therefore, had been stationary for two months.

The examination otherwise was absolutely negative. There was no history of anything suggesting uncinate attacks. Nothing in the examination except her abducens palsy supported the suggestion of a cerebellar lesion given in the history of her complaints.

She remained under observation for a week, during which time she had no special discomforts. The perimetric findings favoured a right temporal lesion, but the diagnosis was uncertain and a choked disc questionable. She was discharged as a "brain tumour suspect" to report for observation.

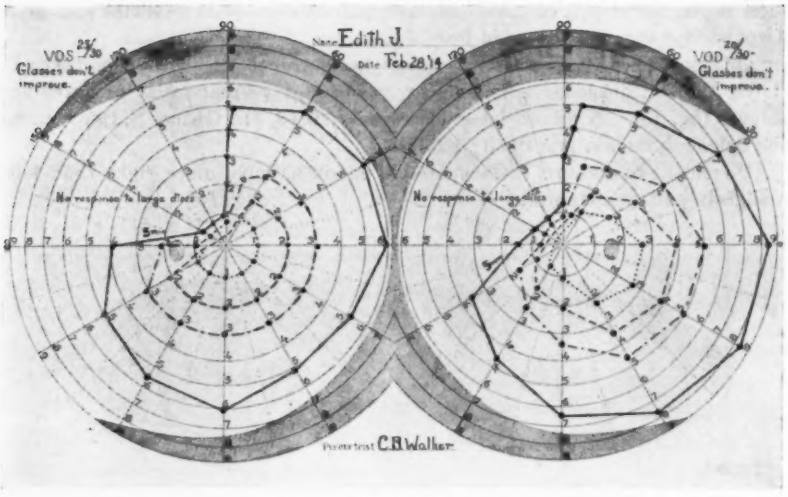


FIG. 14.—*Case* 3 (*contd.*). For comparison with fig. 13 five months later.

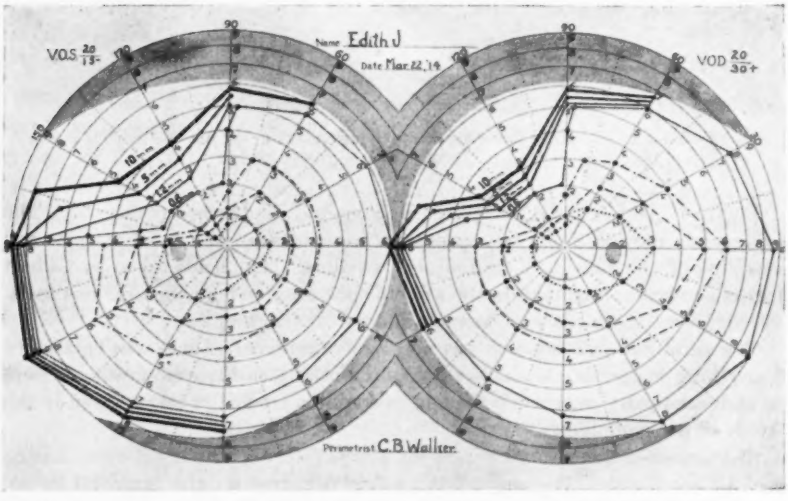


FIG. 15.—*Case* 3 (*contd.*). Fields sixteen days after operation disclosing gliomatous cyst in right temporal lobe.

During the next three months the condition remained unchanged except that her diplopia practically disappeared.

In February of 1914, she had several attacks of severe suboccipital pain with recurrence of the diplopia. This led to her re-entry. The fields were much as before, though the contraction was somewhat more marked (fig. 14). Haziness of the nerve head, however, was definitely more pronounced.

March 6, 1914: Operation. A right subtemporal decompression was performed. The temporal lobe was full, its convolutions flattened and an exploratory puncture through the second temporal convolution disclosed a gliomatous cyst containing about 30 c.c. of straw-coloured fluid.

She was discharged March 24, 1914, her mental condition having greatly improved on the subsidence of her headaches. The fields, as shown by Dr. Walker's charts (fig. 15), had begun to fill out to their normal outlines.

Following her discharge she remained free from symptoms for five years. The abducens palsy disappeared. Though from time to time the decompression area would become prominent and tight, this would soon pass off and there was no real headache. She had been active at social functions, and was engaged in the Y.M.C.A. corps during the war.

On the afternoon of April 16, 1919, while at the dressmaker's, she suddenly became nauseated and vomited. She was readmitted to the hospital three hours later. The decompression area was tense and her vomiting persisted. She began to have auditory hallucinations of people knocking at her door and of telephone conversations. There were visual hallucinations also—of seeing coloured lights and processions of queer figures marching on the ceiling. Her old diplopia again became marked.

Examination showed, as before, nothing but a low grade of papillœdema and a right abducens palsy. The fields of vision, though from inattention they were plotted with some difficulty, showed the same defects as on her first admission (fig. 13).

April 25, 1919: Operation 2. The temporal lobe was again exposed and the cyst was easily entered. It contained about 30 c.c. of straw-coloured fluid which set on standing. The walls were thoroughly fixed with Zenker's fluid until of a leathery consistency and the wound was closed. She made a perfect recovery, and the history records a recession of the field defects (the charts unfortunately have been lost). She was discharged May 22, 1919.

She reported in September, 1919, having been free from symptoms since her discharge. At this time the fields (fig. 16) still showed a slight notching. At the present time, two years from her last operation, and eight years since the onset of symptoms, she remains perfectly well, and her last perimetric examination, April 21, 1921, shows normal field peripheries (fig. 17).

Comment: This experience might be multiplied many times. The defect had never advanced to a hemianopsia but remained quadrantal, and on the side of the lesion, as is often the case, was a little in advance of the other. It involved chiefly the upper field quadrants, and hence indicated pressure on the ventral fibres of the pathway.

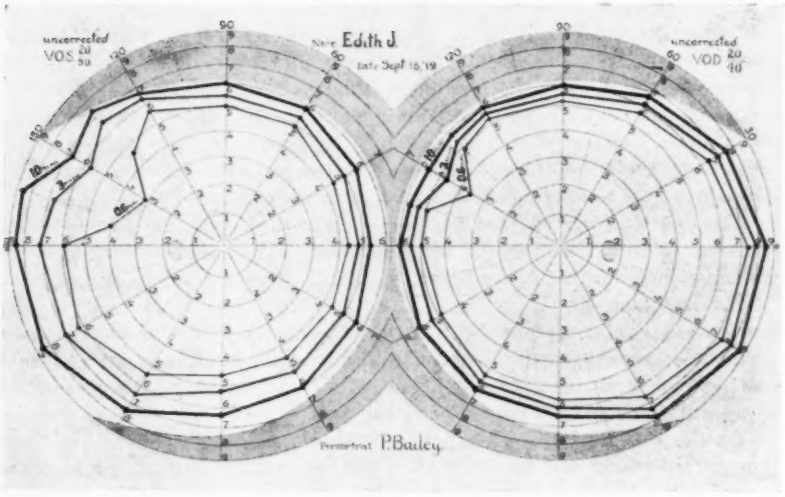


FIG. 16.—*Case* 3 (*contd.*). Showing persistence of slight homonymous defect five months after second operation.

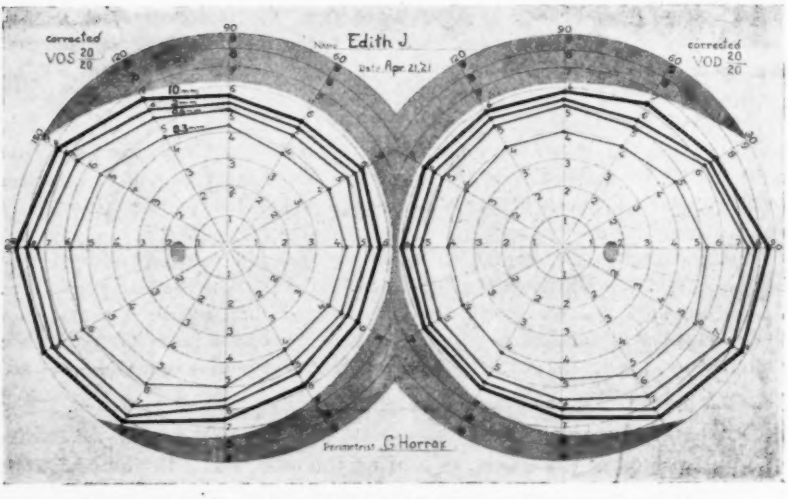


FIG. 17.—*Case* 3 (*contd.*). Normal fields two years after second operation.

Another example of this same type may be given, also a glioma, though one with a less favourable ultimate outcome. The patient was admitted at a time when Dr. Walker was making very detailed studies, as shown by the elaborateness of the first chart (fig. 18), which gives not only the form peripheries, but those for colours as well. It will be seen that there was a homonymous hemianopsia for colour as well as for form when the minute test objects were used. The condition, therefore, was somewhat more advanced than in Case 3, though the lesion was of the same type and in the same situation. A localizing diagnosis would not have been possible without the fields. They show still better than those of the foregoing case that the quadrantal defect on the side of the lesion, as shown by the larger discs, is somewhat in advance of that on the opposite side.

Case 4.—*Homonymous hemianopsia produced by a gliomatous cyst of right temporal lobe. Operation followed by improvement.* (P. B. B. H., Surg. No. 4550.)

April 8, 1916: Admission of Robert S., aged 36, a machinist, referred from the Boston Psychopathic Hospital, with the complaint of headaches and failing vision.

Clinical story: Four years before, he had been struck by a heavy piece of metal on the right forehead and rendered unconscious. There were no subsequent symptoms. For the past year he has not felt well, and has been drinking heavily. Six months before entrance (October, 1915) he had a general convulsion without warning. Since then there have been two similar attacks (November, 1915, and January, 1916). Since December he has had spells of suboccipital headache accompanied by nausea and vomiting. These have increased in intensity and there is now continued discomfort. For two months past there has been progressive blurring of vision. He has had numbness of arms and legs, some unsteadiness of gait, and also tinnitus in the left ear for the past year. He has had a few attacks suggesting *petit mal* in which he feels dazed and experiences a sensation of numbness and pricking in the right arm. Otherwise there has been nothing suggestive of uncinate attacks. He has been treated all this time for stomach trouble.

Physical examination. This disclosed nothing except the following few positive neurological findings. Bilateral choked disc of 5 D. with abundant hæmorrhages. Definite nystagmoid jerks on looking to the left. Slight impairment of hearing, i.e., conduction to watch less good on left than right (6 in. to 24 in.). Positive Romberg with falling to the right.

Aside from the evidences of a marked increase in intracranial tension the findings therefore were few. There was nothing to justify a localizing diagnosis, though it is evident from the history that the first examiner wavered between the suggestive cerebellar signs and the history of possible involvement of the right arm in the *petit mal* attacks. These possibilities were promptly set aside when the fields of vision came to be taken. They showed

a left homonymous defect which therefore lateralized the lesion in the right hemisphere beyond any diagnostic doubt (fig. 18).

April 19, 1916: Operation. Right subtemporal decompression. This disclosed an exceedingly tense temporal lobe. An exploratory trocar was inserted in the second temporal convolution toward the ventricle and at a depth of 3 cm. tapped a large gliomatous cyst containing over 30 c.c. of fluid. The tension was completely relieved. The cyst was widely opened by an incision through the second temporal convolution but its walls were not treated. A fragment of the wall removed for histological study showed a diffuse glioma.

He made a perfect recovery (figs. 19-20), and was discharged May 5, practically without symptoms, though the choked disc had not as yet com-

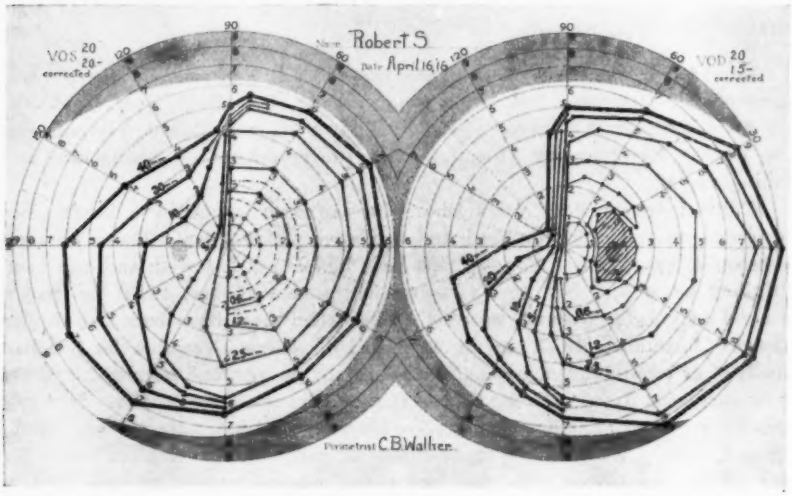


FIG. 18.—*Case* 4. Field taken before operation disclosing a right temporal gliomatous cyst. Note complete hemianopsia to discs below 1·2 mm. size. Also greater advance in defect to larger discs for right than left eye.

pletely subsided. The fields, except those for colour, had by this time partly regained their normal outlines (fig. 21).

December 22, 1916: Reported in person, having been working at his trade since shortly after his discharge. Condition excellent. No symptoms. Decompression soft. Fundi normal. Fields are normal except for a slight defect in the left temporal region to the smallest visible disc (fig. 22).[1]

[1] This patient re-entered the hospital October 22, 1918. Unfortunately no fields were taken at the time as the staff was mostly in army service. He had been quite well until the month previous and had worked regularly until a few days before. His decompression had become tense. The cyst was tapped on several occasions, but immediately refilled. An exploratory operation was performed which revealed an extensive gliomatous growth. He was discharged and his death was reported a few months later.

FIGS. 19 and 20.—*Case* 4 (*contd.*). Condition on discharge sixteen days after subtemporal decompression, showing slight protrusion at seat of defect.

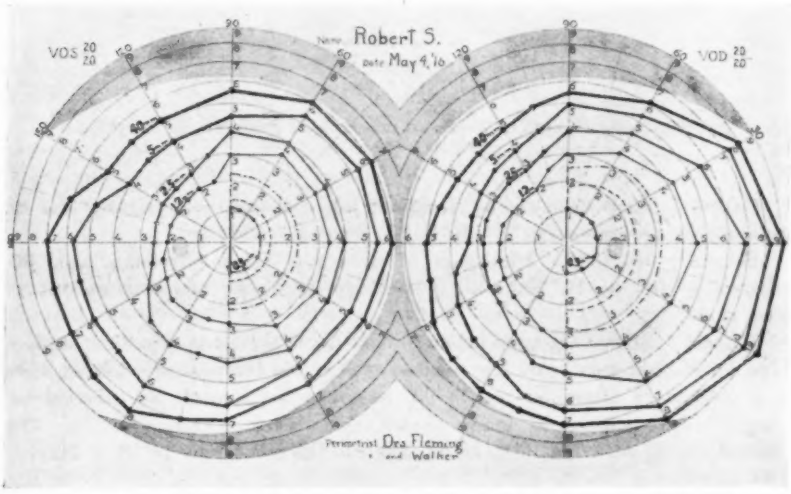


FIG. 21.—*Case* 4 (*contd.*). Fields on patient's discharge sixteen days after operation. Note persistence of homonymous hemianopsia to 0·3 mm. disc.

The following case, owing to the nature of the growth, was far less favourable than any of the preceding ones. The diagnosis of a temporal lesion was sufficiently clear owing to his full-blown uncinate seizures, though without the perimeter the lateralization of the tumour would have been difficult. At the time of his first admission, as will be seen, the fields were regarded as practically normal, though to a minute disc a notch in the right upper quadrant, the forerunner of a more extensive defect, was already apparent.

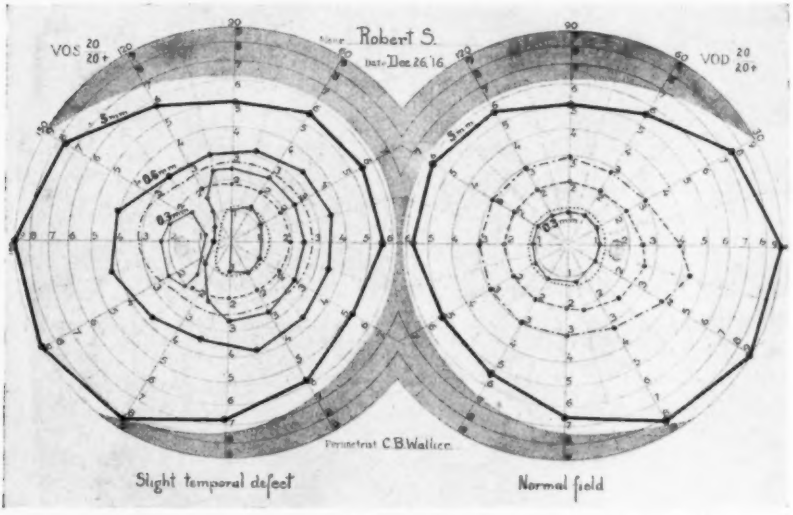


FIG. 22.—*Case* 4 (*contd.*). Fields six months after operation, for comparison with figs. 18 and 21.

Case 5.—*Solid glioma of left temporal lobe with uncinate seizures and an upper right quadrantopsia.* (P. B. B. H., Surg. No. 6273.)

February 16, 1917: Admission of Robert A. C., a physician, aged 36, referred by Dr. A. B. Kanavel, of Chicago, with the major complaint of *petit mal.*

History: Somewhat apprehensive about his health ever since his student days, he for fourteen years has regularly taken his temperature and in view of a customary afternoon rise to 99° has regarded himself as tuberculous. Frequent examinations by many physicians failed to corroborate this. The account of his *petit mal* attacks as taken in the records by Dr. S. J. Harvey is as follows:—

Uncinate attacks: "Eight and a half years ago began to have attacks of *petit mal.* The first one came on suddenly while taking a needle from a

patient's hand. He became dizzy, felt faint, had 'air hunger,' went to a window, opened it and soon revived. One month later there was a similar attack and soon they came with increasing frequency, often ten or twenty a day. The attacks are usually accompanied by what he describes as an exaggeration of his special sense impressions, i.e., flowers become extraordinarily beautiful, odours are intensified, his vision appears to be remarkably keen, so that objects seem to be increased in size. The olfactory hallucinations were never of an unpleasant nature.

"There has never been any loss of consciousness and the attacks have always been so fleeting that a bystander would hardly know he was having one. There is usually an aura 'as if something were to happen,' and during the attack he feigns to be meditating or cogitating over what may have just been said. He states that no two of the attacks are exactly alike but that they vary according to the time of day, his surroundings or the season of the year. For the past two or three years the attacks have been a little more severe; they would leave him momentarily dazed and disorientated; for instance, while talking to a friend he might be seized with an attack which would cause him to stop his conversation, and when he 'came to' would have to ask what he had been saying, or at other times would plead an important engagement to cover his confusion. He has occasionally had a sense of seeing objects to his right which were not there. For some years he has taken bromides in large doses, 70 to 80 gr. per diem."

Headaches: For fourteen months he has had occasional severe suboccipital headaches but without vomiting. During these periods there has been some retraction and rigidity of the neck.

Vision: Has never noticed any diminution of vision, but he had glasses fitted when his headaches came on and found he had been seeing double without being aware of it. For a month he has had considerable bilateral tinnitus.

Physical examination: A robust individual over six feet in height and weighing 230 lb. The findings were absolutely negative except for a low grade of choked disc with swelling of 1 D., a fairly well marked lateral nystagmus, a slight weakness of the right face on expressional movements (discounted by the patient as a personal characteristic) and a slight weakness of the left abducens.

The fields of vision taken at this time with only two test objects (fig. 23) showed to a small 0·3 mm. disc a notch in the right upper quadrant of the left eye, whereas the peripheries to a 5 mm. disc were normal. No special significance was attached to these fields.

It is evident from this early history that the case despite the uncinate seizures was regarded, on the basis of the suboccipital headache and the nystagmus, as a cerebellar tumour suspect. He returned to his home and re-entered the hospital a month later. At this time his choked disc had become more pronounced with elevation of 3 D.

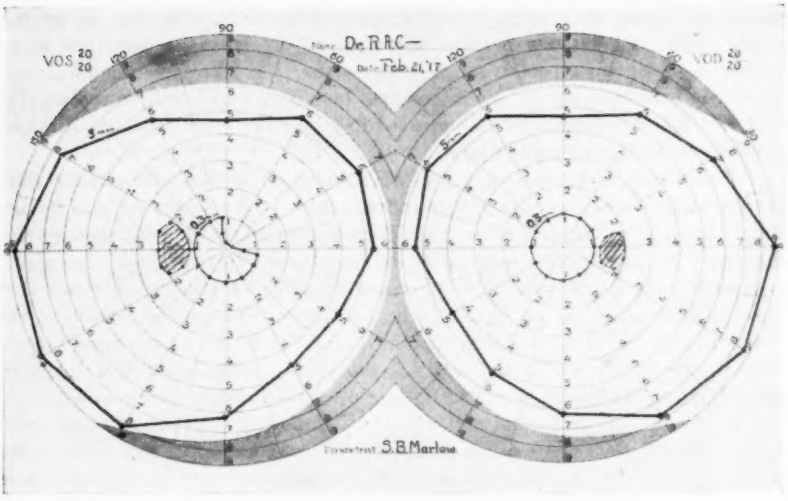


FIG. 23.—*Case* 5. Fields three weeks before operation. Note upper nasal notch in left to 0·3 mm. disc. Not regarded at time as of particular significance.

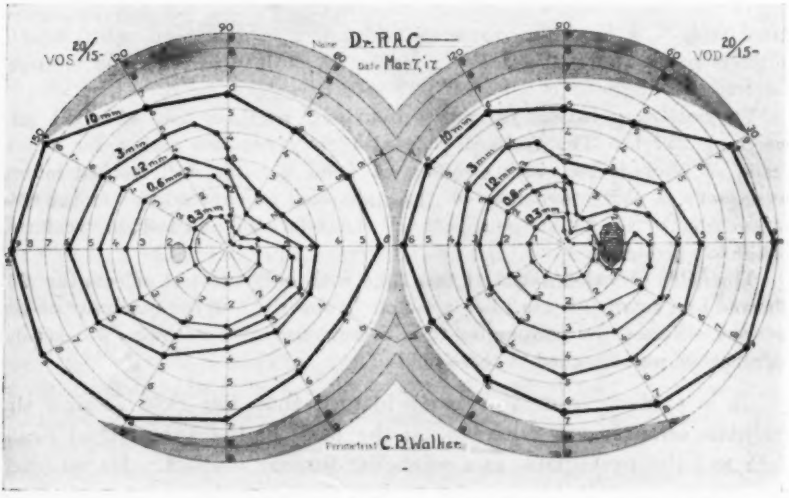


FIG. 24.—*Case* 5 (*contd.*). Fields showing homonymous involvement of which those in fig. 23 gave promise. Note notch at this session to the 0·3 mm. disc in each eye.

and the fields taken by Dr. Walker (fig. 24) showed an advancing defect, the right field now being involved. There remained no doubt as to the situation of the lesion in the left temporal lobe.

March 10, 1917: Operation. Left subtemporal decompression disclosing a tense bulging temporal lobe with flattened convolutions. Exploratory puncture secured about 2 c.c. of yellowish fluid. Diagnosis: glioma.

Subsequent notes: He made a good recovery. The choked disc subsided. He was discharged March 24. During my absence in the Army he reported by letter on October 10 as being in good health despite his continued *petit mal* attacks. He is reported to have died January 8, 1918. It may be assumed that before the end he had a complete homonymous hemianopsia.

The following case illustrates the terminal condition of complete hemianopsia which in the preceding would probably have been observed had a perimetric test been made toward the end. The patient had visual hallucinations referred to the defective fields (left) and a few gustatory impressions suggesting uncinate stimulation, though there were no "dreamy states." The lesion, therefore, even without the field defects, might possibly have been localized in one or the other temporal lobe, but the characteristic fields made its lateralization possible.

Case 6.—*A rapidly growing cystic glioma of lower right temporal lobe producing upper quadrantopsia unrelieved by operation and advancing to hemianopsia.* (P. B. B. H., Surg. No. 4002.)

December 14, 1915: Admission of Jacob B., aged 11, a right-handed boy, with the complaint of headaches and vomiting.

Clinical history (notes by Dr. Horrax): "Onset a year before admission, with frontal headaches and pain in the eyes, chiefly the right. The headaches were usually associated with nausea, vomiting, and often with dizziness. These symptoms have increased until of late headache has been constant. For a month he has had buzzing in the right ear, and he recently was operated upon in a local hospital for adenoids. Some occasional blurring of vision has also been observed during the past month, but his acuity remains unimpaired. For the past week there has been some double vision, and on three occasions he had had definite visual hallucinations, always to the left. Once he had the impression of seeing a boy on the wall. Another time a man dressed in white was seen sitting by the fire bending over to tie his shoe, and this impression remained a long time. One night, after his admission, he thought he saw some children sitting around a desk on the wall, but they were gone when the nurse turned on the light.

"On one occasion, just before entrance, he thought that he had smelled and tasted peaches when none were around, and after his hospital admission he insisted that he smelled and tasted roasted peanuts. These two occasions

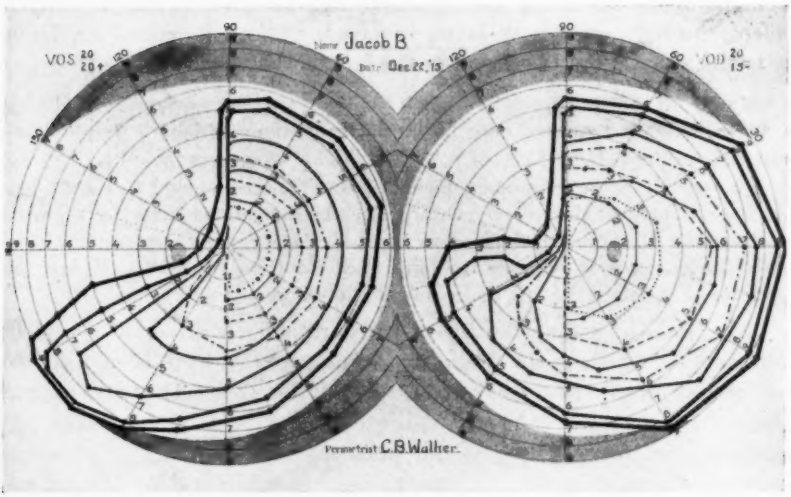
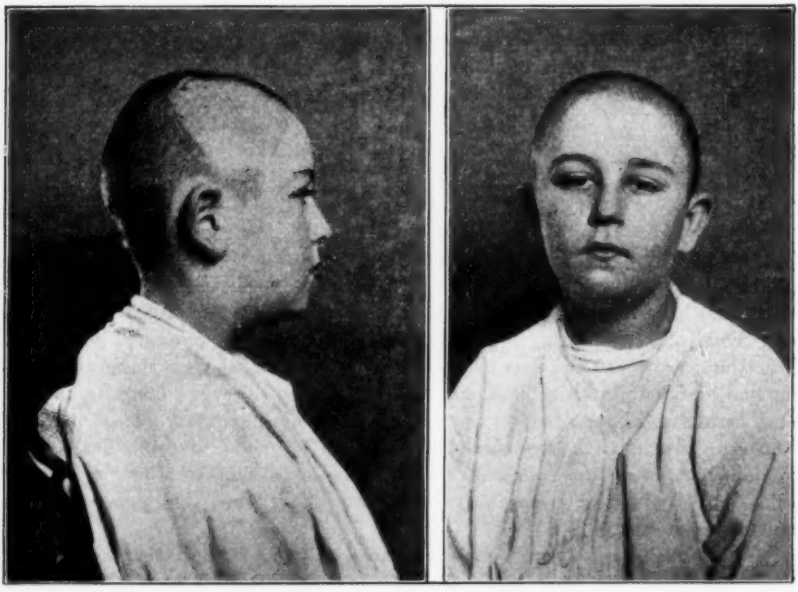


FIG. 25.—*Case* 6. Fields shortly before operation disclosing a right temporal glioma.



FIGS. 26 and 27.—*Case* 6 (*contd.*). Two weeks after operation showing situation of bone flap and slight temporal protrusion through the subtemporal defect.

were not associated with anything resembling a 'dreamy state.' He had never had any known losses of consciousness."

Physical examination: A healthy, intelligent, and co-operative boy, with no positive neurological findings except for a choked disc of 3 D., signs of intracranial pressure shown by the X-ray, and a left upper homonymous quadrantopsia (fig. 25).

December 31, 1915: Operation. The right hemisphere was exposed by an osteoplastic flap carried well down in the temporal region. Here the bone was found greatly thinned. The dura was opened over the temporal lobe and reflected upward. The convolutions were not greatly flattened, nor was the Sylvian fissure pushed upward. Palpation of the lower part of the temporal lobe gave the impression of a subjacent cyst. An exploratory puncture in the second temporal gyrus at a depth of 4 cm. reached a small cyst, giving a few cubic centimetres of typical yellow fluid. A subtemporal bone defect was made and the flap replaced.

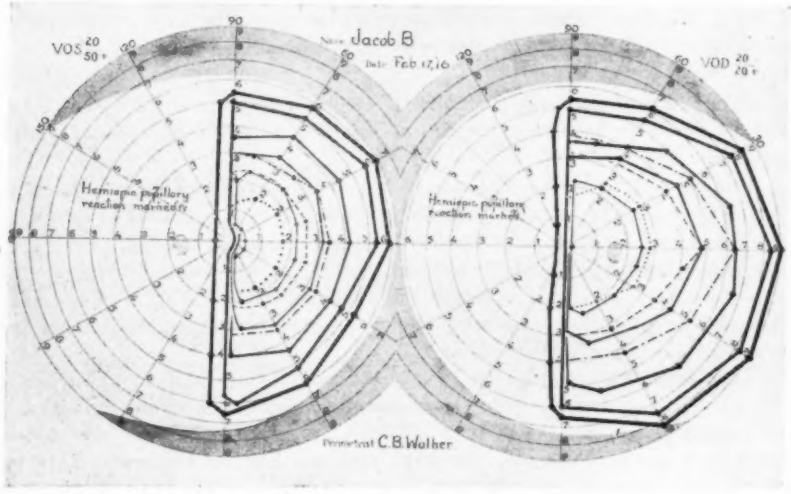


FIG. 28.—*Case* 6 *(contd.)*. Fields two months after operation, for comparison with fig. 25, showing ultimate complete homonymous hemianopsia with macular inclusion.

The boy made a perfect recovery (figs. 26, 27), with relief of all subjective symptoms, except that once during his convalescence he had another vivid hallucination—of seeing "a lady wearing a hat." His choked discs subsided, but the fields of vision remained unaltered.

He was discharged January 22, 1916, much improved, but his period of relief was short. The decompression gradually became tense, and he began to have some left-sided numbness and weakness.

He re-entered the hospital February 16, 1916, owing to the increase of symptoms. His fields at this time (fig. 28) showed a complete hemianopsia. An operation was performed, and a large mass of gliomatous tissue containing small cysts was removed from the temporal lobe. There was again a temporary period of improvement, and he was discharged. The prognosis was hopeless, and he died a few months later at his home.

Doubtless, in a case of this kind, the geniculate body itself comes to be involved in course of time, and in all probability could a series of fields have been taken, there would have been a stage in which the macula was not completely bisected.

The patient whose history follows was first seen at such a stage, and there was considerable subsequent improvement, though in the end a fatal issue. In this case, too, visual hallucinations were a pronounced feature, as the story will relate.

Case 7.—*Cystic glioma of right temporo-sphenoidal lobe, with hallucinations of vision and a lower left quadrantopsia. Operation. Temporary improvement.* (P. B. B. H., Surg. No. 3043.)

June 9, 1915: Admission of Mrs. Helen L., aged 33, referred by Dr. D. B. Steuer, of Cleveland, Ohio, with the complaint of "headache and optic neuritis."

Clinical history: *Petit mal* attacks. In 1911, four years before admission, during the course of her first pregnancy, she began having attacks of *petit mal*. In these attacks her left hand was involved and would feel swollen. They were followed by twitching of the fingers and drawing up of the arm and face. She was invariably conscious during these attacks. After the birth of her baby these seizures, which had occurred many times a day, decreased in number and gradually disappeared completely.

Headache and diplopia: In 1912 she began having headaches referred to the right frontal region. They have become increasingly severe, culminating in a particularly bad attack three months ago, since when they are said to have been less troublesome. In 1912. also, she had for two weeks a period of diplopia. The eye-grounds were examined, and a choked disc of 4 D. was found. This condition continued throughout 1913, during which time she was under the care of a succession of "specialists."

Grand mal attacks: On four occasions she has had a general convulsion with loss of consciousness. The first occurred in June, 1913, the others eighteen, twelve and ten months ago respectively.

Anosmia and hallucinations of smell: Immediately after the birth of her child in 1912, she found that she had lost the sense of smell, and during the past year she has frequently had olfactory hallucinations, occasionally disagreeable ones. She describes them as "like something from another world; nothing that is in the environment—I can't describe it."

Left hemiparesis: First noted two years ago, as dragging of the leg and tendency to drop things from the hand. The symptoms became more definite three months ago in association with the series of severe headaches. The face then became weak and the arm and leg still more so. Some loss of sensation on the left has also been noted.

Failing vision: Choked disc first noted three years ago, but vision not greatly impaired until two months ago. She is conscious of loss of vision to the left.

Hallucinations of vision: Present for the past two or three weeks. For example, the following quotation is taken from the history: "She repeatedly told the nurse on awakening that a woman friend of hers, whom she named,

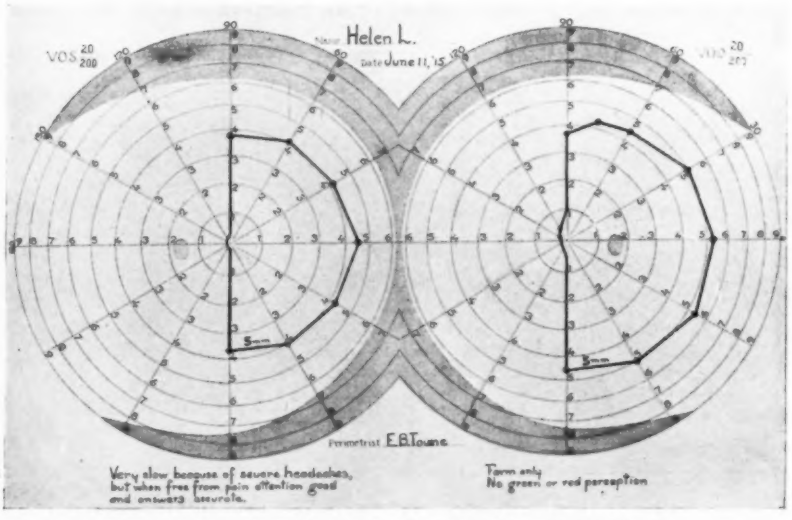


FIG. 29.—*Case* 7. Fields on admission shortly before operation.

was in the room, and she wanted her husband called to talk to this woman. The imaginary person stayed in the room all night, moving about, but did not talk. She was always to the patient's left. When the patient turned her head to follow her, the hallucination would also go to the left and disappear from the field of vision. To-day during the taking of the history she asked several times if the doctor had come back. Finally, she said, 'Well, who is it in the room?' On questioning it appeared that a man with a gold helmet on his head came into the room with the examiner and sat in a chair beside him. During the history-taking he also had a paper and took notes. He did not leave the room until the examiner left. His face was not recognized and she could not describe it clearly. Both examiner and this hallucination were to patient's left side."

Physical examination: A well-developed and nourished woman, showing the following positive neurological signs: a choked disc of 4 D., a total anosmia, a sharply-cut left homonymous hemianopsia (fig. 29). Hemihypæsthesia over entire left side of body with slight weakness of musculature. Deep reflexes increased to exaggeration on the left, with clonus and a positive Babinski.

June 14, 1915: Operation. Right osteoplastic flap, disclosing a tense dura. On exposing the hemisphere the temporal lobe protruded markedly and the Sylvian fissure was much dislocated upward. The convolutions were greatly flattened. A needle introduced into the posterior portion of the first temporal convolution struck a large gliomatous cyst containing nearly 100 c.c. of yellow fluid which clotted on standing. The tension completely subsided. This cyst was at such a depth that no effort was made to make an incision into it nor to treat its walls.

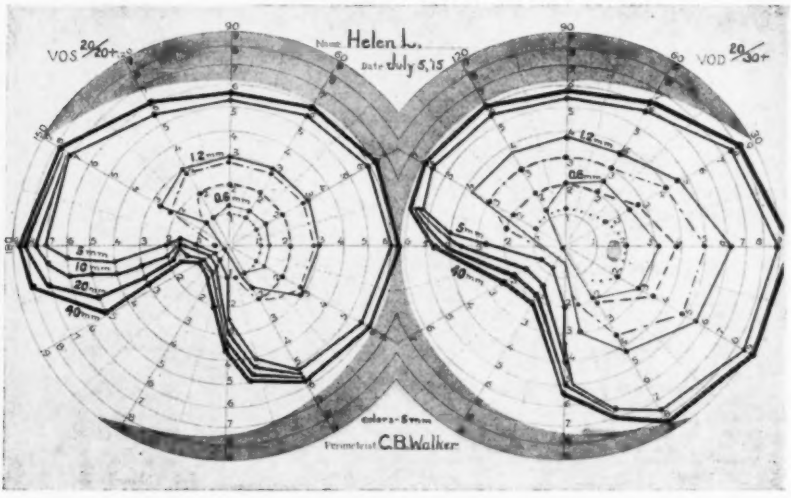
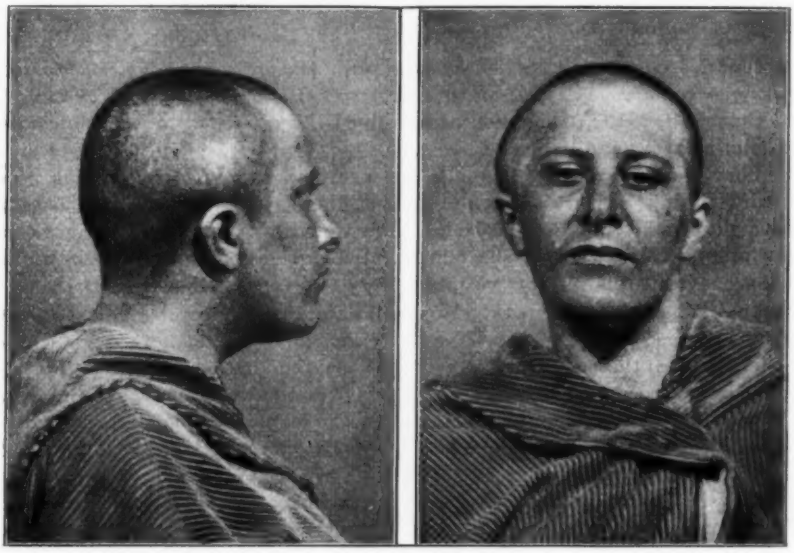


FIG. 30.—*Case* 7 (*contd.*). Fields three weeks after operation which disclosed a high temporal defect (for comparison with fig. 29).

Post-operative: For a few days she continued to have many bizarre visual hallucinations, always seen to the left. They were usually of people doing queer things, for example: "A woman riding horseback with a sheet of music propped in front of her and she used her hands as though playing the piano on the horse's neck." On one or two occasions also complained of disagreeable odours. These attacks subsided, however, and she made an excellent recovery with practical disappearance for the time being of all her previous symptoms. Normal reflexes, sensation and sense of smell were all regained. The choked

discs subsided with unexpectedly good vision in view of their original appearance. The fields taken on July 5, by Dr. Walker (fig. 30), showed a widening of the peripheries to the left with a persisting lower quadrantal defect which the high temporal situation of the lesion would have led one to expect.

She was discharged to her home July 9. Dr. Towne's final note is as follows: "Of late no convulsions, headache, hallucinations of smell, taste or vision. Left side now almost as strong as right, face still a trifle weak; dynamometer right 24, left 18. Vision very much improved—cf. fields. Choked discs almost flat. No Babinski; no ankle clonus. Deep reflex, still +, but less than before. Mental condition markedly improved; still rather jocular but perfectly sane; knows what serious condition she was in at entrance." (Figs. 31, 32.)



FIGS. 31 and 32.—*Case* 7 (*contd.*). Shortly before discharge, showing situation of osteoplastic flap and considerable protrusion through subtemporal defect.

Subsequent note: She passed into other hands and apparently no further fields of vision were taken. The cyst had to be tapped on several occasions. Her death was reported to have occurred a year after her discharge.

Even without the fields there should have been no real difficulty in making a diagnosis in this case, though the early attacks of *petit mal*, said to have begun in the hand, might have led one to anticipate a lesion which originated in the paracentral convolutions. Her homony-

mous hemianopsia, however, was enough to place it below the Sylvian fissure; and that the lower part of the pathway at least was not destroyed is apparent from the fields of July 5, which showed a return of vision in the upper areas.

The combination of visual, gustatory and olfactory hallucinations in a temporal lobe lesion is deserving of comment. The growth was of such a size that it must have occupied a large part of the lobe and, as was noted at the time of the operation, the Sylvian fissure was markedly dislocated, a condition not infrequently observed in large temporal lobe tumours.

The following case, one with a more favourable outcome, is another example of a cyst in practically the same situation as the foregoing, though at the time of admission the defect had not advanced to the stage of hemianopsia.

Case 8.—*Gliomatous cyst of right temporal lobe producing a lower homonymous quadrantal defect. Operation. Recovery.* (P. B. B. H., Surg. No. 11283.)

October 8, 1919: Admission of Dorothy D., aged 19, with the complaint of fainting spells and headaches.

Four years previously (1915) she had a severe fall on the ice, striking her head. To this she attributes her present illness, for soon after she began having headaches. These headaches grew worse, and one day, a year after the injury, while at school she fainted. Subsequently, fainting attacks with slight convulsive movements occurred every two to three weeks.

In 1916, at the Boston City Hospital, a right subtemporal decompression was performed by Dr. E. H. Nichols. Though her discomforts were completely relieved thereby until a few months ago, the fainting attacks continued. They are preceded by a feeling of numbness over the whole body, but there is no gustatory sense impression. The convulsive features of the attacks, hardly apparent at first, have of late become more pronounced. A large subtemporal protrusion has gradually formed (cf. figs. 36, 37).

Physical examination: An intelligent, alert, co-operative young woman, free from discomforts, and in excellent physical condition. While under observation she had a convulsion, beginning with a vague stare. For two or three minntes she could respond to questions and denied the presence of any olfactory or gustatory impressions. The left face, then the arm and leg, began to twitch, and a general convulsion followed.

Positive neurological findings: A bilateral choked disc with beginning secondary atrophy. Nerve head on the right elevated 1·5 D. On the left 3 D. Slight hypæsthesia over the left side of body without appreciable motor involvement, though the deep reflexes were possibly more brisk on the left than right. The fields of vision revealed a lower left homonymous defect (fig. 33).

October 10, 1919: Operation. An osteoplastic exploration was made to expose fully the right hemisphere. The temporal lobe was bulging, and its

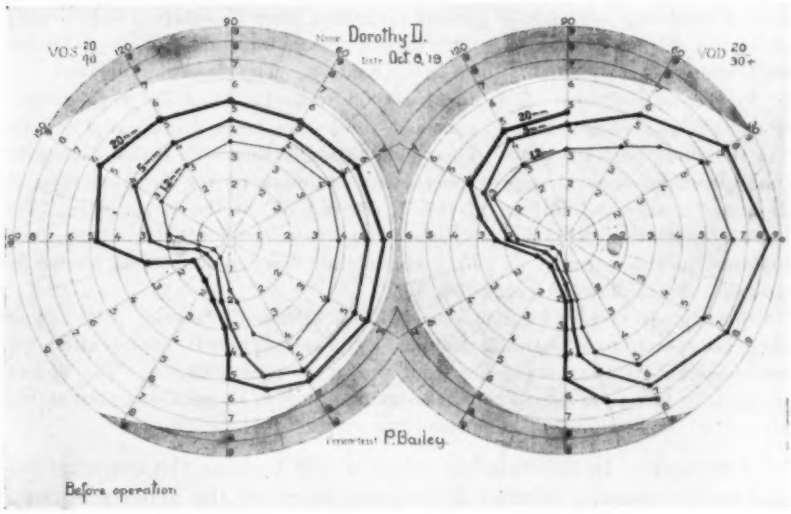


FIG. 33.—*Case* 8. Fields before operation which disclosed a high temporal lobe cyst.

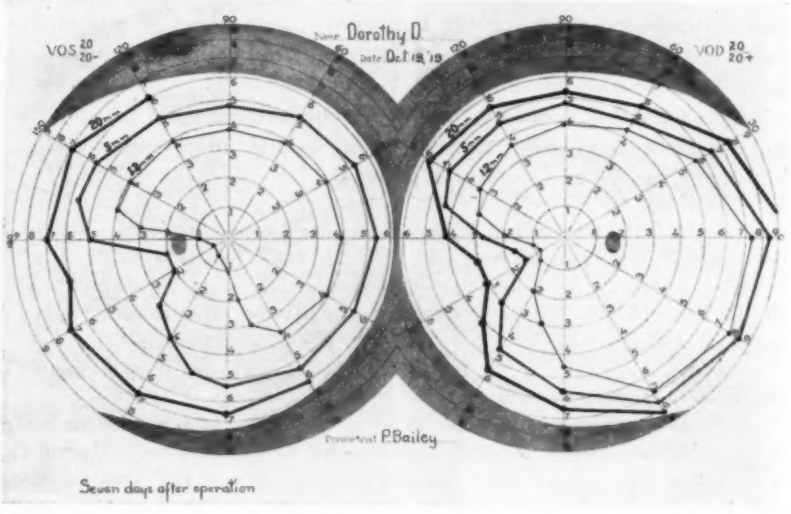


FIG. 34.—*Case* 8 (*contd.*). Fields nine days after operation, showing recession of defect.

convolutions flattened. Puncture of the first temporal convolution revealed a large cyst containing characteristic yellow clotting fluid. The cyst was widely opened disclosing a nodule of tumour projecting from its external wall directly under the old decompression. This nodule was completely excised. The cyst wall was treated as usual with a tissue fixative. The flap was replaced.

Post-operative note: She made a good recovery; had no more convulsions; the choked disc rapidly subsided, the hypæsthesia disappeared and she has remained perfectly well. The fields taken October 19, seven days after the operation, showed improvement, with widening of the peripheries to all discs and a definite filling in of the defect (fig. 34) to the larger ones. Ten days later (October 29), the day before her discharge, the field outlines were restored to the normal (fig. 35), where they have remained as shown by subsequent examinations (figs. 36, 37).

Subsequent note: She had one fainting attack Christmas, 1919. Since then no symptoms of her old trouble. Her most recent examination was made March 16, 1921, six years from the onset of symptoms. The decompression is now quite flat. Fields are normal. Fundus shows slight perivascular streaking: no swelling. She is steadily at work.

Comment: In this case, therefore, a cyst high in the temporal lobe and rather posterior exerted its pressure effect on the upper portions of the pathway, leaving the lower fibres unimpaired. The rapid restoration of the field peripheries show that it was a pressure effect only. There was nothing in her neurological examination to suggest a temporal lesion, except the perimetric findings, and in their absence as likely as not the temporal lobe might not have been exposed in the exploration. A puncture of the lobe at the first operation probably would have revealed the lesion.

The following history is one of an incurable glioma in which the field defects were seen to pass from a mild involvement through advancing stages to complete blindness.

Case 9.—*Right temporal diffuse glioma with uncinate seizures. Successive operations. Advancing process.* (P. B. B. H., Surg. No. 3774.)

October 28, 1915: Admission of Mrs. Sadie G., aged 28, referred by Dr. H. B. Eaton of the Boston Dispensary, complaining of *petit mal*, headaches and loss of vision.

Present illness: Dr. E. B. Towne's recital of her story may be quoted in full from the hospital record.

"*Petit mal* attacks: Began about a year ago. She says a peculiar feeling runs suddenly through the body, lasting only a few seconds. During this time her arms and legs feel numb and relaxed. She is told that she stares and that her face becomes flushed, and later pale. This happens possibly once a week. They are liable to come on while she is at the table. There is

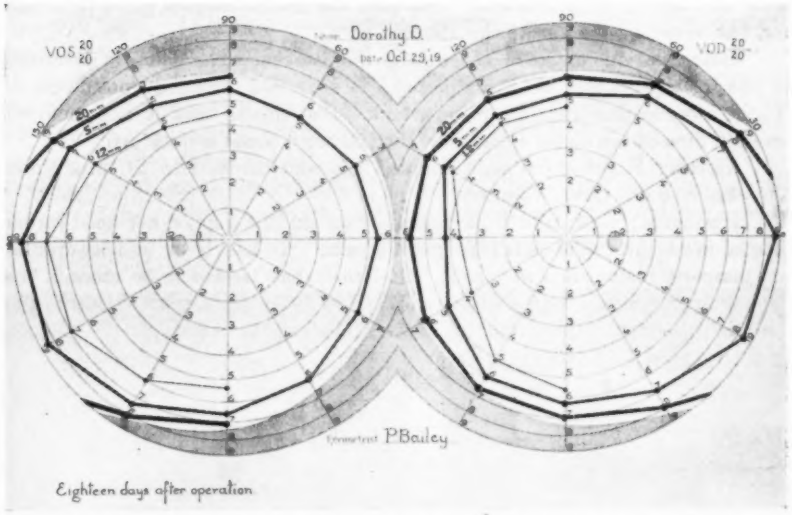


FIG. 35.—*Case* 8 (*contd.*). Showing normal peripheries on discharge nineteen days after operation, for comparison with figs. 33 and 34.

FIGS. 36 and 37.—*Case* 8 (*contd.*). *Circa* two weeks after operation, showing the position of the bone flap with soft protrusion at the seat of the old subtemporal defect, the scar of which is apparent and which directly overlay the tumour nodule.

never any loss of consciousness. She says she knows what is going on about her, but never speaks during the attacks.

"Hallucinations of smell accompany the attacks. The odour is described as like fresh paint and is so distinct she is surprised others do not detect it. It goes away immediately after the attack. She thinks that it has occurred possibly nine to ten times altogether, the last time about a week ago.

"Nausea: Began about eight months ago and comes two to three times a week. She has rarely vomited.

"Headache: She says it is really a pain on the top of her head rather than a headache. She calls it a 'pulling pain.' It has been in both frontal and temporal regions and in vertex. Of late it has tended to be more in the right temporal region than anywhere else. She is some days entirely free and again is miserable for two to three days. Headache is liable to be brought on by any sudden motion, as getting out of bed or sitting down suddenly?"

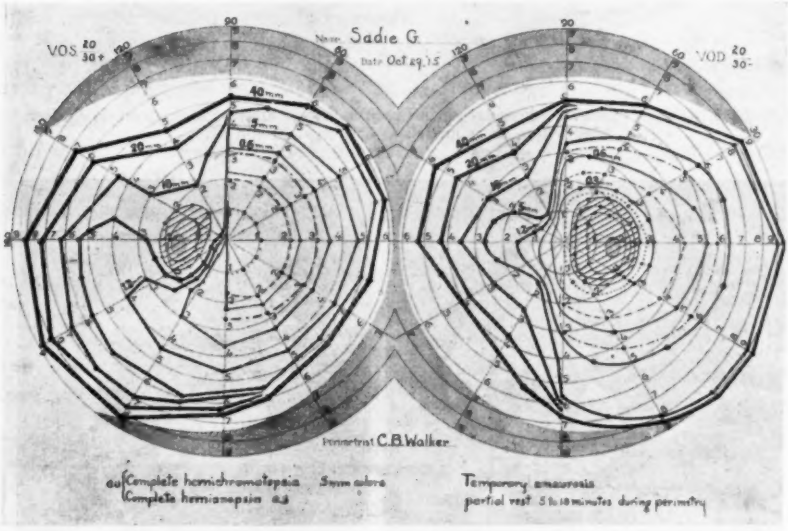


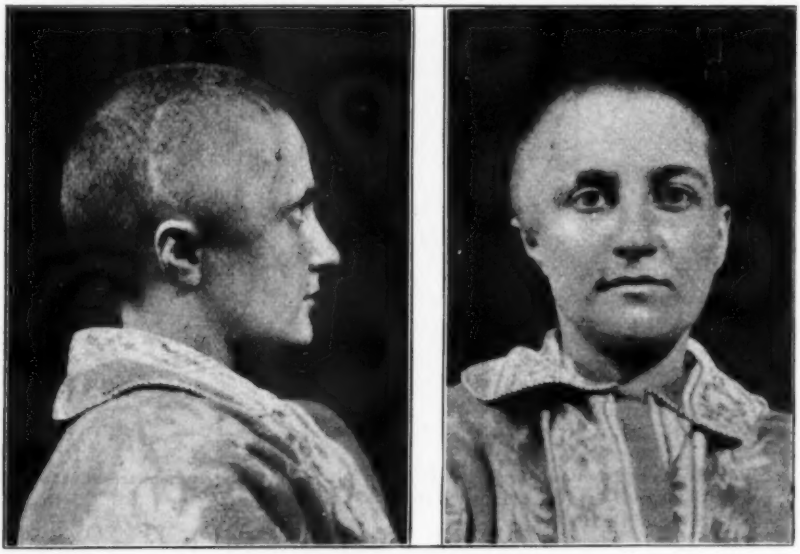
FIG. 38.—*Case* 9. Fields shortly before operation showing left homonymous hemianopsia complete in both eyes to a 0·3-mm. disc. Charts show unusually well gradations of discernible defect to larger discs.

Loss of vision: Four weeks ago she noticed that newspaper print was blurred and that she could read only with difficulty. Thinks that eyes were equally affected and that they have become gradually worse. At present cannot read print in telephone book. There have been attacks of temporary amblyopia quite frequently during the last two weeks.

Twitching of left face: During the past four weeks it has happened three

to four times. Corner of mouth draws up and eye closes. It is difficult to open eye again. It lasts a few moments. There is no twitching of arm and leg. No association with *petit mal*.

Hallucinations of vision: One week ago was walking across the room and thought that she saw a cat to her left. Says cat was very black and was hunched up as if angry. She turned around and looked about the room before she was convinced that the cat was not there. Two days later she again saw the same black cat on the left side. On two to three other occasions she has seen what she describes as black specks to her left, and she turns her head to look at them. There have been no other visual hallucinations.



FIGS. 39 and 40.—*Case* 9 (*contd.*). Patient on discharge, to show situation of osteoplastic exploration with subsequent protrusion through subtemporal defect.

Physical condition: A healthy, vigorous, intelligent, co-operative patient, sound in every respect. She was right-handed.

Neurological examination: This revealed absolutely no abnormalities except an advanced choked disc with hæmorrhage and exudates, and an elevation of 5 D. right and 6 D. left.

Her history was sufficient to suggest a temporal lobe lesion, but the side could not have been determined without the perimeter. An abnormality in the fields was first detected by Dr. Woodward, the interne, and they were subsequently plotted in great detail by C. B. Walker (fig 38). They showed an oncoming left homonymous hemianopsia with the process more advanced

in the right eye. To the 5-mm. discs there was a hemiachromatopsia which showed for form likewise when the 0·3-mm. test objects were used.

November 5, 1915: Operation. A low osteoplastic resection was made over the right hemisphere. The bone in the temporal region was thinned by pressure. A subtemporal defect was made to allow for subsequent decompression. The temporal lobe was full and bulging. Aspiration revealed no cyst. An incision was made through the first temporal convolution and carried down to a depth of about 3 cm., when it came upon a soft reddish tumour. A fragment removed for verification showed glioma.

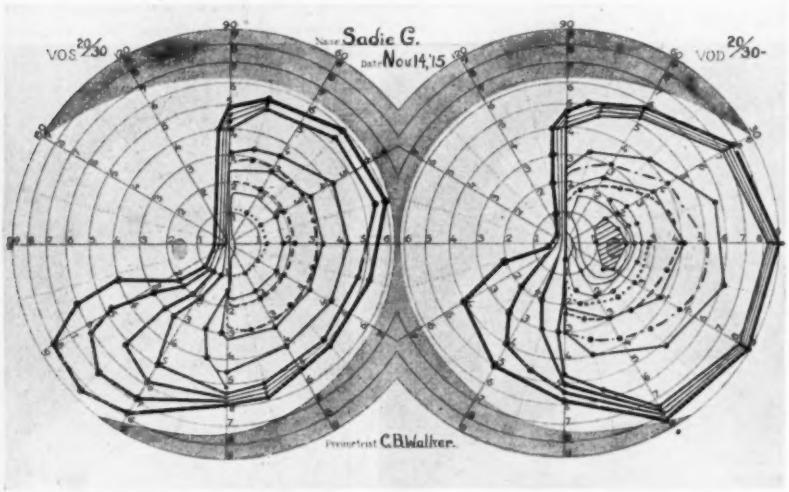


FIG. 41.—*Case* 9 (*contd.*). Fields three weeks after operation, for comparison with fig. 38, to show increasing field defects.

Post-operative: She did exceedingly well (figs. 39, 40). Headache and *petit mal* attacks ceased. The choked disc had subsided to 1 D. by November 26, the day of her discharge, The fields, however, showed a slight advance in the process (fig. 41).

In January, 1916, she reported subjectively free from symptoms. Dr. Walker's fields at this time (fig. 42) show a further slight advance in the process.

She remained perfectly well for nearly four years despite an increasing subtemporal protrusion. A note on January 24, 1917, states: "Patient reports in excellent condition. No symptoms. Says she never felt better in her life. There is a fairly large, rather soft subtemporal protrusion which she conceals perfectly with the arrangement of her hair (fig. 43). No further hallucinations or *petit mal* attacks. No fields taken."

March 10, 1919: She reported on my return from abroad in excellent general condition. No headache, hallucinations or *petit mal* attacks. The

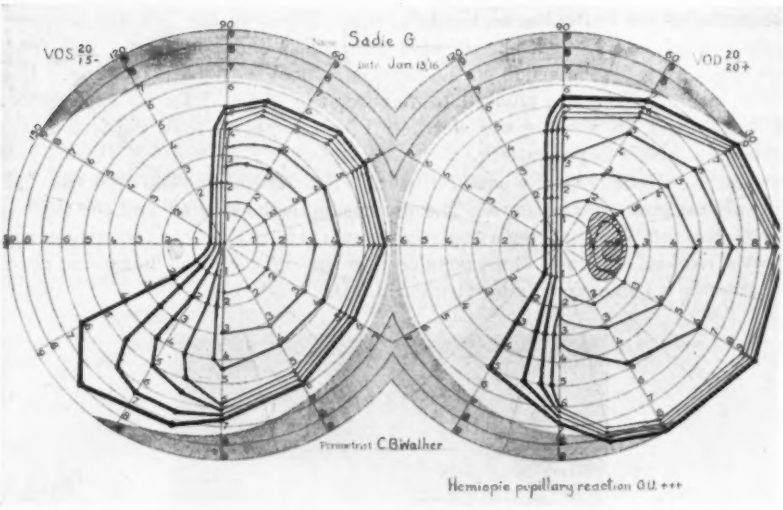


FIG. 42.—*Case* 9 (*contd.*). Field two months after operation to show further advance. (Cf. figs. 38 and 41.)

FIG. 43.—*Case* 9 (*contd.*). Patient fifteen months after operation, when she was subjectively free from symptoms.

protrusion, however, has been growing steadily larger and she has had occasional slight twitching of the left face. Vision in her left eye has been perfect until lately, and she is able to read without glasses. But from secondary atrophy the vision in the right is reduced to light perception. (No fields taken.) She was advised to re-enter the hospital for an exploration. It was hoped, in view of the slowness of the process, that the lesion had undergone cystic degeneration.

March 21, 1919: Re-entry. (Surg. No. 10165.) The process had advanced during the ten days. The protrusion had enlarged and the twitching of the left face had been replaced by a palsy (fig. 44). The vision was greatly reduced, shadows alone were seen in the right eye and in the left there was merely a hemiopic tubular vision (fig. 45).

FIG. 44.—*Case* 9 (*contd.*). Patient before last operation, showing huge protrusion with left facial palsy.

April 11, 1919: Secondary operation. A soft tumour mass the size of a fist practically extruded itself through the subtemporal defect on exposing the brain. As could be seen, this represented only the external part of the growth. Closure.

This procedure gave but little subjective relief and led to no improvement in vision. She died February 18, 1920, six years from the onset of her symptoms.

The patient's history on admission suggested a temporal lobe lesion in view of her visual and olfactory hallucinations. There was nothing

in the examination, however, to show whether the process was right or left, unless the slightly more advanced choking of the left disc could have been taken to favour this side. The visual fields on rough finger tests showed no hemianopsia, so that without the perimeter it would not have been possible to lateralize the process. The perimetric defects which were disclosed were typical of a low right temporal lesion affecting the ventral bundle of the radiation. As is usual the eye on the side of the lesion showed, throughout, the more advanced defect.

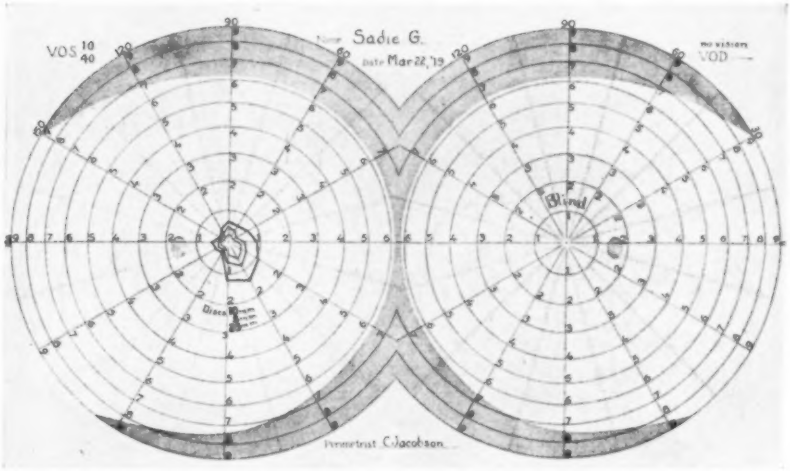


FIG 45.—*Case* 9 (*contd.*). Residual of vision on last admission four years following her first operation. For comparison with figs. 38, 41 and 42.

This peculiarity of an unequal progress in the defect on the two sides is still better shown in the next and last of the ten cases which have been selected for this report. The diagnosis was most obscure until the field defects began to appear.

Case 10.—*Brain tumour suspect. Decompression. Tumour localization made probable by early field distortions. Partial enucleation of meningeal endothelioma of left temporal fossa.* (P. B. B. H., Surg. No. 10671.)

June 19, 1919. Admission of Mrs. Ruth K., referred by Dr. E. O. Morrow, of Canton, Ohio, with the complaint of headache, vomiting and defective vision.

History: One year before entrance received a blow on the head and was temporarily stunned. Five months later began having suboccipital headaches, also pain over the left eye. Soon the eye began to protrude and turn inward.

Other complaints were transient numbness of left forehead, and pains referred to the left teeth. Morning nausea and vomiting frequent of late. Considerable dizziness and general weakness. Occasional subjective numbness in fingers of right hand.

Neurological examination: The only positive findings were a bilateral choked disc; slight exophthalmos of the left eye; left abducens palsy; left pupil larger than right: slight expressional weakness of right face. A presumptive diagnosis of tumour involving the temporal lobe was made, possibly an endothelioma of the Gasserian envelopes. This was further supported by the disclosure, on taking her fields, of a notch in the upper nasal quadrant in the left eye (fig. 46).

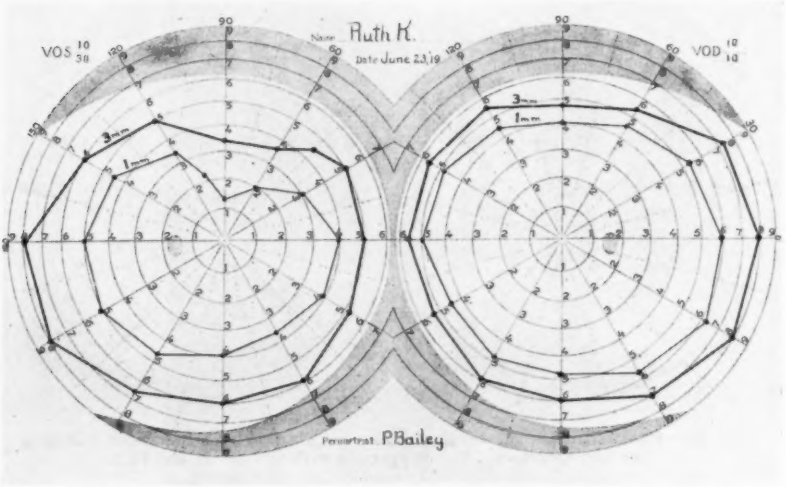


FIG. 46.—*Case* 10. Fields on admission of patient who was a left temporal lobe tumour suspect.

June 25, 1919: Operation. Left osteoplastic flap. Combined exploration and decompression. The brain was found under practically normal tension. The ventricle was tapped to permit of a good view under the temporal lobe in the direction of the fifth nerve. Negative findings. The lobe seemed a little full but an exploratory needle introduced in two places revealed no lesion. Closure.

The symptoms remained unaltered. Occasional periods of numbness in the fingers of the right hand continued, accompanied sometimes by slight paraphasia. On July 7 she complained for the first time of sudden gustatory sensations—like peppermint. On this date the fields of vision showed a marked advance in the defect in the left eye and a homonymous notch in the right similar to that first seen in the left (fig. 47).

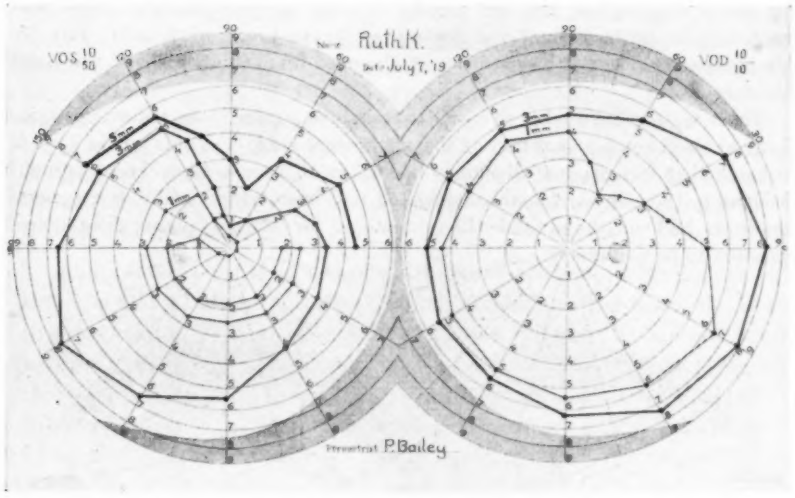


FIG. 47.—*Case* 10 (*contd.*). Field two weeks after negative exploration showing advance in process. (Cf. fig. 46.) On July 16 there was found a slightly more marked defect in the fields.

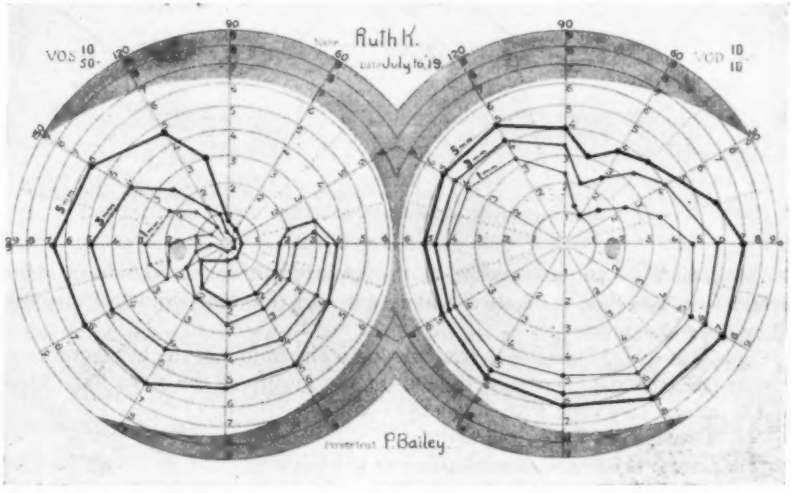


FIG. 48.—*Case* 10 (*contd.*). Fields three weeks after operation, showing further advance in defect.

On July 16 there was found a still more marked defect in the fields (fig. 48). The choked disc had largely subsided by this time and there was practically no protrusion of the decompression area. A week later, July 23, this peculiar field defect was still more advanced (fig. 49) and here it remained stationary.

There were grave doubts as to what course to pursue. An antero-posterior X-ray showed some absorption of the sphenoidal ridge and this made it seem probable that our original diagnosis had after all been correct and a tumour situated farther forward than anticipated had been overlooked. Her general condition had improved and the symptoms for the advancing field defect appeared to be stationary.

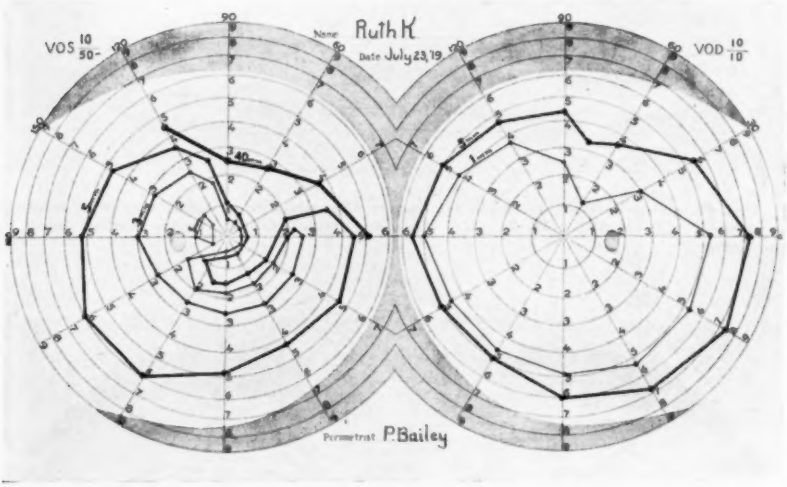


FIG. 49.—*Case* 10 (*contd.*). Fields one month after negative exploration, showing continued progress in the defect, chiefly of the left eye.

Most of the symptoms were so inconspicuous that no particular stress would have been laid upon them had it not been for the perimetric changes. They were in summary :—

Subjective : (1) Occasional numbness and discomfort of the left teeth.

(2) Occasional momentary tingling of the fingers of the right hand associated on two occasions with slight aphasia.

(3) Fleeting gustatory impressions.

Objective : (1) Slight exophthalmos of the left eye.

(2) Slight choking of left disc more than right.

(3) Upper right homonymous field defects.

August 11, 1919 : Operation 2. Partial enucleation of a large endothe-

lioma. A more thorough and more anterior exploration was carried out at this time. The margin of the tumour was brought into view. It had been covered and concealed by a thin shell of the temporal lobe. Its base of attachment was obviously at the sphenoidal ridge. The large growth was fragmented in removal and in the attempt to complete the enucleation there was bleeding and the remaining fragment had to be left in position. Even so, a cavernous sinus thrombosis was provoked which led to such a degree of proptosis that the eye, owing to keratitis, was finally enucleated on September 4. Normal field outlines were regained in the right eye by the time of her discharge.

One cannot be sure in a case of this kind (with a tumour compressing the lobe and a tumour, moreover, which arises from the meninges in such a situation that the optic tract itself may possibly have been pressed upon) whether the field distortion was actually due to involvement of the temporal loop. It, however, is more than likely, but in either case the diagnosis of a temporal lobe lesion was chiefly based on the advancing quadrantal defect. For the purposes of this paper, the case is important for the reason that the original fields of June 23 represent the earliest stage of such a defect that has been recorded in the series. To make a diagnosis of temporal lobe tumour largely on the basis of a notch in the upper nasal field in one eye would seem absurd were it not for the accumulated evidence regarding the localizing significance of these partial defects which this paper is intended to emphasize.

Temporal Lesions other than Tumour.

Though this communication deals primarily with tumour cases, it is not to be forgotten that the same principles hold true for lesions of other sorts. The illustration given at the outset, of a gunshot wound of the temporal lobe, is a case in point, and in all probability when the war material has been fully worked up, important contributions to our knowledge of temporal lobe symptomatology will be forthcoming, even though the urgency of military service in most hospitals for the recently wounded may have precluded such details as the use of the perimeter.

That the position of the radiation is poorly understood even when the perimeter has been used and a hemianopsia disclosed is evident enough, and in the minds of many physicians a homonymous hemianopsia indicates an occipital lobe lesion. I have even known of an occipital exploration for a shrapnel ball on this basis, conducted under the direction of a neurologist, when, as subsequent events showed, the missile actually was in the temporal lobe. Probably all of us have made

similar mistakes in tumour cases when the distractions of war could not be offered as an excuse.

But in civil life there are other sources of injury to the temporal lobe far more common than gunshot or stab wounds which may produce these field defects. Fractures of the base of the skull are possibly the most frequent source, for, as is well known, they are prone severely to contuse the tips of the temporal lobes. There are a number of cases of this sort in our records in which the perimeter has been employed with the disclosure of sharply cut sector-shaped defects (fig. 50). And I am

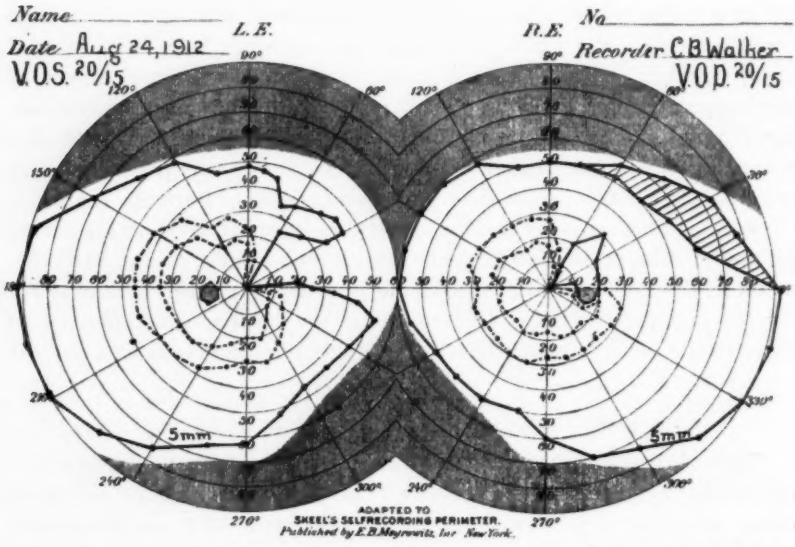


FIG. 50.—Sharply angulated homonymous field defects reaching to the central point following fracture of base of skull, with contusion by contrecoup of left temporal lobe.

under the impression that these traumatic lesions are more apt to produce defects in the fields characterized by sharp angulations than are tumours in whose presence the defects are ordinarily, if not always, rounded off. I have twice seen such defects as the result of the so-called prize-fighter's fracture of the base following a blow on the jaw transmitted through the condyle (fig. 51).

Another and still more important condition producing these defects is a temporal lobe abscess. Difficulty is often experienced in differentiating between a possible cerebellar or temporal lobe lesion when patients

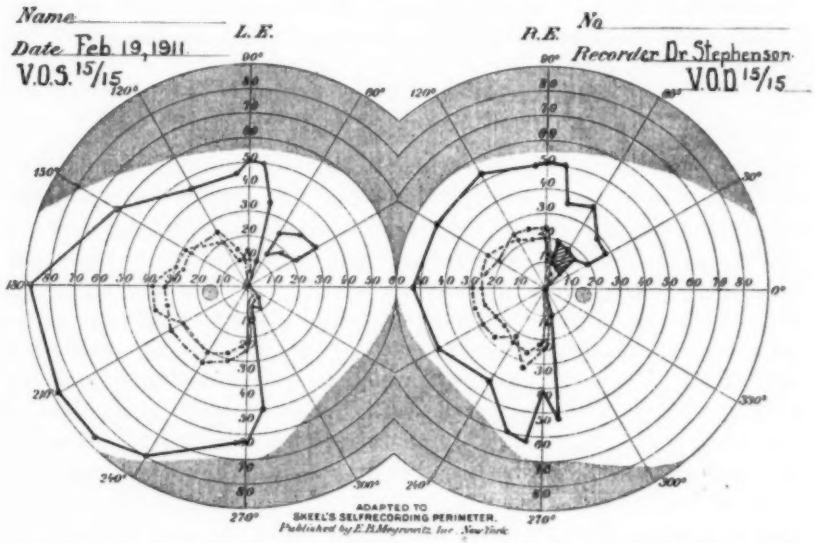


FIG. 51.—Incomplete homonymous hemianopsia following left temporal lobe lesion following a blow received in a boxing contest.

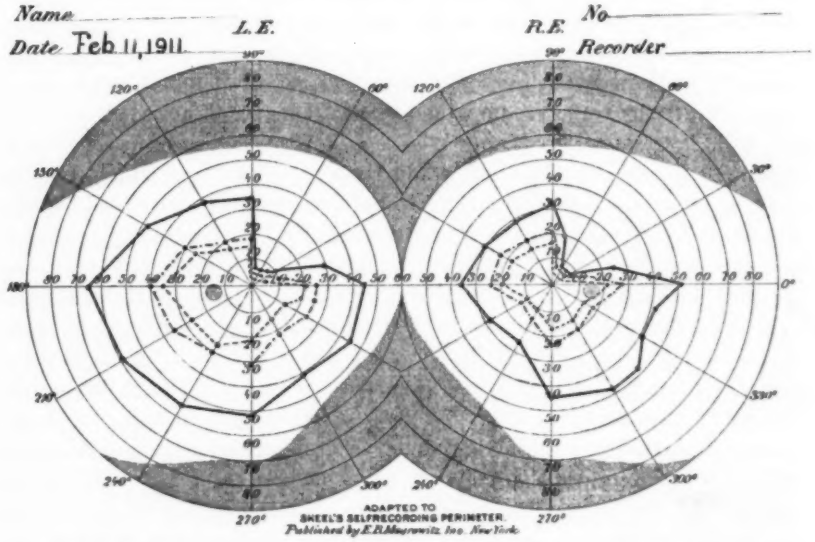


FIG. 52.—Right upper quadrantopsia in patient with left temporal lobe abscess.

are under the suspicion of having an abscess secondary to an otitis media. The perimeter, under these circumstances, may give invaluable information (fig. 52).

The General Symptomatology of Temporal Tumours.

It is but a scant twenty years since Byrom Bramwell, in an article on the localization of intracranial tumours [2], made the statement that tumours of the temporo-sphenoidal lobe and more particularly those of the right side were of all tumours the most difficult to diagnose and to locate, because these regions represented the most silent area of the brain. This statement by an eminent neurologist long interested in the special topic of brain tumours was utilized twelve years later by Foster Kennedy to introduce his article, which to the present time remains perhaps the most important single contribution (certainly in English) to the subject [4]. After referring to the comparative rarity of temporo-sphenoidal tumours Kennedy made a careful symptomatic analysis of nine verified cases from the records of the National Hospital at Queen Square. In conclusion he gave what he considered to be the essential symptom complex of these tumours for both right and left lobes. Common to a tumour of either lobe were: (1) convulsions of major and minor (uncinate) type, (2) bilateral choked disc, usually more marked on the side of the lesion, (3) post-epileptic transitory disturbances of motion and of the reflexes which later become persistent. In addition to these symptoms alike for both sides, a tumour on the left was said usually to be accompanied by some degree of aphasia.

It is evident from Kennedy's report that he was dealing with tumours of a more advanced grade than most of those which I have used as examples, for by the time an aphasia occurs in left-sided lesions together with weakness in the opposite face and marked change in the reflexes there is, in my opinion, considerable involvement of areas beyond the confines of the temporal lobe. It is, however, but natural, owing to the great advances in neurological surgery during the past decade, that many more of these lesions are to-day identified and at a much earlier stage than at the time of his paper. Hence in a considerable number of our verified cases, as indicated in some of those used in illustration (Cases 2 and 3 for example), the symptom complex pictured by Kennedy had not as yet appeared and the localization depended largely upon the field defects. The possibility of co-existent field defects was in Kennedy's mind, but he dismissed it with the following statement:—

"An examination of the pathological findings in my patients suggests the probability of hemianopia or hemiachromatopsia having been present during life owing to the proximity of the growth to the optic tract and radiations. In all cases the existence of hemianopic defect was considered and negatived as the result of examination. Five patients were examined perimetrically for various colours; the sole abnormality discovered was the concentric contraction of the visual field so often associated with severe, and especially with protracted papillœdema."

This quotation is given for the purpose of laying further emphasis on the value of the field defects in question, in the early diagnosis of temporal lobe lesions, rather than with any intent of pointing out an oversight of my distinguished predecessor.[1]

A tabulation of the symptoms recorded in the histories of the 59 verified temporal lobe tumours in my series has been prepared. As heretofore stated, in 6 only of the 39 cases in which the patient's condition made reliable perimetry possible were the fields normal at the time the tests were made. These were with two exceptions temporal endotheliomata which did not greatly deform the lobe and which in the absence of a choked disc probably would not have been detected at all had it not been for the unilateral exophthalmos and the overlying cranial hyperostosis, the usual tell-tale of these not uncommon tumours. The exceptions were: (1) a meningeal angioma; and (2) a glioma in which the recorded normality of the fields is perhaps subject to question. The fact, however, that field defects were present in 33 of 39 cases tested indicates that the perimeter, as a diagnostic aid in these lesions, is possibly the most important agent of all.

Though it was not my intent, in outlining this paper, to enter into a general discussion of temporal lobe symptomatology, the table which has been prepared to aid in this study brings out certain things in relation to the diagnosis which deserve to be briefly commented upon.

Generalized convulsions.—These were recorded in 20 of the 59 cases. They usually were few in number. A single convulsion without aura or sudden loss of consciousness, possibly unattended by convulsion, had been the inaugural symptom in three or four instances. A few of the patients had had only two or three convulsions and twelve was the largest recorded number.

[1] While this paper was going through the press my attention was called to an important article, "Die Tumoren des Schläfenlappens," by Albert Knapp, in the *Deutsche Zeitschr. f. d. g. Neurol. u. Psychiat.*, 1918, **43**, 226-89. In this elaborate study based on two cases Knapp mentions the fact that hemianopsia has been described. He does not lay any great stress upon this condition however.

The so-called uncinate seizures.—For the forty-five years since Hughlings Jackson's original description of these peculiar disturbances they have held the ground as the most distinctive and characteristic of all symptoms relating to the environment of the temporal lobe. This notable contribution to cerebral symptomatology, entirely consistent with the characteristics of its discoverer, was based on knowledge gained through animal experimentation subsequently transferred to the clinic and ultimately confirmed at the post-mortem table.

In the 59 cases of this series attacks often described as *petit mal*, or as more or less characteristic "dreamy states," were present in 24 cases. In 14 only of these cases, however, were there any olfactory or gustatory impressions and in some of these the possible uncinate character of the attacks was not particularly definite and appears to have been read into the patient's history after the lesion had been verified as a temporal one by operation. Nevertheless, when these Jacksonian seizures do occur in outspoken fashion they are unmistakable and of great diagnostic value, but, even so, speaking for the temporal lobe as a whole, they are far less common than are the field defects. Indeed, as might be expected of a lesion involving the uncinate gyrus, field defects were present with but one exception in every case having presumed uncinate seizures in which the use of the perimeter was not precluded by blindness or some other cause. On the other hand, field defects were present in eighteen cases in which no history suggestive of *petit mal* or dreamy states could be secured.[1]

Visual hallucinations.—Examples of these states have been given in the case histories (e.g., Case 6). Considerably to my surprise these visual pictures, often as vivid impressions, are recorded in thirteen out of the fifty-nine cases in the series. They, presumably, are part and parcel of the uncinate seizure, for in only three of the thirteen cases did they occur unrelated to what were regarded as attacks of this type. Hughlings Jackson, with Beevor, was among the first to describe this symptom [3] in a patient who had the impression of seeing "a little black woman engaged in cooking." A good example of the same thing is given by Kennedy in the history of his first case. These projection

[1] It is a matter of surprise to me that Hughlings Jackson did not emphasize the importance of the perimeter as well as of the ophthalmoscope, on which he laid such great stress in his writings (*Brain*, 1915, **38**, p. 391). The latter is unquestionably the more important instrument of the two, and as the profession was loath to adopt it, and still is for that matter, he may have felt that it was enough to urge its employment without forcing the perimeter on a reluctant profession at the same time.

pictures cannot be at all uncommon, and whether they are related in any way to the olfactory and gustatory discharges of the typical uncinate fit or are merely associated with some pressure implication of the visual pathway or the geniculate body, I am at a loss to say. Certainly they bear some relation to the damaged geniculo-calcarine radiation, for in this series, whenever its situation has been mentioned, the hallucination has always been referred by the patient to the side opposite that occupied by the lesion, in other words, in the defective fields.

One of the most remarkable examples of these hallucinations of which I have record occurred in one of the patients in my Baltimore series. It was a young man under observation for a long period who had repeated uncinate attacks, many of which were observed and described in detail. They invariably had the same character and left a vivid and disagreeable recollection. As in a dream he saw, always to his left side, his father and some other men in an unfamiliar room engaged in a game of cards which ultimately led to an altercation. The scene would fade and leave him with a bad taste and smell of something indescribably horrid. Objectively, during the course of ordinary conversation, possibly while standing, a vague look would come over his face, he would turn his head and stare to the left and shortly after would begin to make smacking and tasting movements with his lips. This was followed by an expression of disgust and the attack would be over. He had an homonymous upper quadrantal defect produced by a temporal lobe glioma.

One would naturally expect visual hallucinations to be a feature of occipital rather than of temporal lobe tumours, but the former are far less common in my series and, though they have not been thoroughly studied with this point in view, it is my impression that they are less prone to have *petit mal* attacks and that the subjective visual phenomena, if any, are more likely to be of colours and lights than of pictured scenes.[1]

[1] Though I have not fully looked up the general subject of visual hallucinations in this connection, it appears to me that those who have observed these states in association with a homonymous hemianopsia have taken it for granted, without any real justification, that the lesion necessarily was in the occipital lobe. This certainly is true of the case reported by Paul Camus (*L'Encéphale*, 1911, **6**, Part I, 521-531) and also of some of the other cases in the literature to which he refers. In an earlier paper by Pick (*Amer. J. M. Sc.*, 1904, **127**, 82-92) there is a good example of what I would take to be a left temporal tumour which he localizes, however, in the angular gyrus. All that can be said of such case reports is that without a post-mortem examination or verification of the situation of the lesion at operation, judging from the cases in this series, one at least is not justified in ascribing too much to the occipital lobe.

Auditory symptoms.—In view of the supposed relation of the temporal lobe and particularly of the transverse temporal gyrus to audition and the speech mechanism, it is extraordinary how slight and inconspicuous are the clinical evidences of any disturbance in this sphere. Certainly in none of the cases in this series has there been anything suggestive of auditory hallucinations of musical sounds or imagined speech, as might have been expected.

Without wishing to provoke a discussion regarding the location of the centres for the perception of speech, it has always been a matter difficult for me to understand why after a left subtemporal decompression for an unlocalizable tumour no disturbance of hearing occurs even though the lobe may bulge markedly through the defect. On the analogy of the paralyses which are known to result from the protrusion of cortex through a defect made over other parts of the brain, one would certainly expect some disturbance of hearing from a protrusion of the first temporal gyrus if this is as important an area as we have been led to believe.

In this series of 59 cases hearing was absolutely unimpaired in 38 of the patients. In eleven cases there was tinnitus heard in one or both ears but it was never a particularly constant or annoying symptom as in acoustic tumours. Slight contralateral deafness was recorded in five cases, slight relative deafness on the side of the lesion in two cases. In the remaining cases the tests were obscured by an old otitis media. To be sure, these tests were made merely by the voice, the watch and the fork and without any such accuracy of measurement as the perimeter gives for the fields of vision, but certainly, were contralateral deafness a common sequence of temporal tumours, occasional cases with pronounced loss of hearing would be encountered, and of these the records contain no example.

So far as aphasia is concerned there can be little question but that, when in left-sided cases a disturbance of speech is evident, one almost invariably can detect some evidence at the same time of a right facial weakness which indicates that the pressure effects of the tumour are exerting themselves on areas above the Sylvian fissure. I am a little in doubt, after re-reading Kennedy's paper, whether he regards the aphasia sometimes seen with left-sided temporal tumours as really temporal in origin. To my thinking, however valuable it may be in tumour lateralization, aphasia is a neighbourhood symptom and not a true temporal lobe phenomenon.

The temporal versus a cerebellar syndrome.—It would seem improbable that one could ever be misled by the symptoms of a temporal

lobe tumour into making the diagnosis of a subtentorial lesion. This nevertheless may occur, particularly in the absence of uncinate seizures and when field defects either are wanting or the perimeter cannot be used.

In fifteen of the fifty-nine cases there was definite nystagmus and in some of the patients it was a marked feature. Dizziness and vertigo are not unusual, and curiously enough primary suboccipital headaches have been fairly common. Then, too, in a few cases there has been definite ataxia and marked static instability. A few years ago one of my assistants, who had been devoting himself particularly to the cerebellar symptomatology and was an expert in the interpretation of Bárány tests, was led to make a suboccipital exploration in a patient who did not survive, and autopsy disclosed a temporal lobe tumour. By a sad oversight the fields of vision in this case had not been taken, for the cerebellar symptoms were supposed to be sufficiently definite to make this unnecessary.

It is not at all infrequent for my assistants or myself to make a tentative rating of a patient on the first superficial study as a cerebellar suspect and to have this presumptive diagnosis ruled out beyond question as soon as the fields have been taken. The chief difficulties arise when the patient is blind or uncooperative. It is of course one of the traditions of neuro-surgery that one may easily mistake a frontal for a cerebellar case or vice versa. This is well brought out by the tables of operations given in Tooth's studies of the National Hospital series, and I have twice been led to make a cerebral exploration for tumours as definitely localizable in the long run as are those of the acoustic nerve. Undoubtedly one must also bear in mind the possibility of confusing a temporal and cerebellar lesion, and it is safe to say that in every case of any obscurity whatsoever the perimeter should be employed.[1]

[1] In his article, "Die Tumoren des Schläfenlappens," by Albert Knapp (*op. cit.*), emphasis is also laid upon the difficulty of diagnosis between a temporal lobe and a cerebellar lesion. In his summary he says: "In the third place, of especial importance are the disturbances of equilibrium which are likely to be chiefly confused with cerebellar symptoms, and can be described as a pseudo-cerebellar temporal lobe ataxia. Less often there are other symptoms which are characteristic of affections of the posterior fossa, such as pain and stiffness in the neck, grinding of the teeth, corneal areflexa, nystagmus, and abducens palsy." He further adds that if these three things—oculomotor palsy, pressure against the cerebral peduncle, and inco-ordination—are present, the diagnosis of a temporal lobe tumour is assured. He evidently regards these three things as the essential feature of temporal tumours and the other symptoms of secondary importance, even disturbances of taste and smell.

Summary and Conclusion.

(1) The temporal lobe is a common seat of cerebral tumour (fifty-nine cases in a series of 276 verified supratentorial tumours).

(2) In the fifty-nine verified temporal lobe tumours, perimetry, owing to the advanced stage of the process, was precluded in twenty cases, but of the remaining thirty-nine homonymous field defects indicating involvement of the temporal loop of the optic radiation were present in thirty-three instances.

(3) Heretofore the most important symptom for temporal lobe localization has been the occurrence of the so-called uncinate fits, but in this series, even including as such all attacks of *petit mal* without gustatory impression, they have been recorded in twenty-four cases only.

(4) Visual hallucinations have been a frequent symptom of the temporal tumours in this series (thirteen out of the fifty-nine cases).

(5) Auditory phenomena in this series are conspicuous by their absence. In a few cases only has there been tinnitus and rarely some lowering of sound perception.

(6) The chief errors of diagnosis arise (i) when, with a total median hemianopsia, the occipital lobe is considered to be the tumour seat; (ii) in the absence of demonstrable field defects, when symptoms supposedly of cerebellar origin are to the fore.

Hence it is fair to conclude that perimetry gives us information of paramount diagnostic value, particularly in the early recognition of temporal lobe tumours, the partial field defects short of a hemianopsia being especially characteristic of involvement of the optic radiation in this region.

References.

[1] Archambault, La Salle. "Inferior Longitudinal Bundle and the Geniculo-calcarine Fasciculus," *Albany Med. Ann.*, January, 1909.

[2] Bramwell, Byrom. *Brain*, 1899, **22**, 37.

[3] Jackson, Hughlings. *Ibid.*, 1889-90, **12**, p. 358.

[4] Kennedy, Foster. "The Symptomatology of Temporosphenoidal Tumours," *Arch. Int. Med.*, 1911, **8**, 317-50.

[5] Meyer, Adolf. "The Connections of the Occipital Lobes and the Present Status of the Cerebral Visual Affections," *Trans. Assoc. Amer. Physicians*, 1907.

[6] Walker, C. B. "Colour Interlacing and Perimetry," *Trans. Amer. Ophthal. Soc.*, 1916, **14**, 684.

[7] *Idem.* "Quantitative Perimetry: Practical Devices and Errors," *Arch. Ophthal.*, 1917, **46**, 537-61.

[8] *Idem.* "Neurological Perimetry," *Trans. Ophth. Sect., Journ. Amer. Med. Assoc.*, June, 1917, 189-202.

REFLEX MOVEMENTS AND POSTURAL REACTIONS IN QUADRIPLEGIA AND HEMIPLEGIA, WITH ESPECIAL REFERENCE TO THOSE OF THE UPPER LIMB.

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INTRODUCTION.

SHERRINGTON'S original studies of the reflex reactions of the hind limbs in decerebrate and spinal animal preparations failed to excite much clinical interest until Babinski drew attention to the diagnostic significance of the plantar reflex in man.

This clinical discovery was indirectly responsible for a closer study of Sherrington's researches by neurologists, with the result that the reflex activities of the lower limbs in man have been the subject of many and important investigations. These have shown that the movement evoked by plantar stimulation is not an isolated phenomenon but forms a part of a general flexion or withdrawal action of the lower extremity. Its analogy with the phenomena described by Sherrington in sub-human movements under similar conditions and its presence in the healthy human infant gave it phylogenetic and ontogenetic significance.

The impetus given by these researches has led to widespread results. The questions of spasticity and other reflex reactions, as they occur in hemiplegia and paraplegia, have been reconsidered in the light of physiological experiments and much theoretical and practical knowledge has been gained in regard to the reflex behaviour of the lower limbs under normal and abnormal conditions.

Desultory attention only has hitherto been given to analogous reactions in the upper limb, largely owing to the rarity of cases of spastic quadriplegia of spinal origin.

Our interest in this subject was first excited in 1916 by the observation of a nociceptive reflex in the upper limb of a patient suffering from a high cervical injury and we have been fortunate enough to have had under our care five examples of quadriplegia showing reactions of this kind. Not unnaturally a search was made for similar phenomena in patients suffering from hemiplegia of cerebral origin and reactions presenting analogous features were frequently found.

It was soon apparent that the effects of stimulation of an upper or lower limb in both quadriplegia and hemiplegia were not confined to the stimulated part, but, under favourable conditions, were expressed as a generalized response in the somatic musculature.

Remote results were not found in response to artificial stimulation only but were evoked by strong voluntary efforts of the patients to make movements against resistance. A prolonged study of these widespread reactions has, we believe, thrown light on the whole question of the so-called "associated movements" which have long been recognized in connection with hemiplegic states.

The nature of muscular tone as a postural reaction had of necessity to be considered in relation to these problems, and numerous observations were made on our patients from this point of view.

As an introduction to our subject there will be described typical examples of quadriplegia and hemiplegia with analysis of the phenomena

they presented. Later chapters are devoted to a consideration of the relation of these phenomena to reflex reactions in decerebrate animals and to the theoretical conclusions which appear to be justified.

An appendix contains summarized records of some of the more important cases on which our paper is based.

Chapter I.—Description of an Example of Spastic Quadriplegia.

Case No. 1.—Charles V., aged 35, late Sergeant, Leicestershire Yeomanry, was wounded in the neck by a spent bullet on February 7, 1915. He was standing in a trench, leaning with his back against the parapet and talking to a sentry when he felt "something like a brick" hit him at the back of his head. He dropped immediately into the trench. Although dazed he remained conscious and heard the sentry call out "Sergeant V. is wounded." He remembers being placed on a stretcher and carried out of the trench. He was unable to speak for some time after he was hit and had considerable difficulty in breathing, being compelled to take quick, short breaths. He retains impressions of only a few disconnected incidents on his journey to the dressing station. According to his statement he was completely insensitive and paralysed, except for movements of his diaphragm, in his trunk and upper and lower extremities. Reflex micturition became established soon after his arrival at the dressing station and a catheter was passed once only, two days later, in the hospital at Boulogne. He arrived at 13th Stationary Hospital, Boulogne, on February 9, and we are indebted to Lieut.-Colonel Gordon Holmes for a note which he made on the patient's condition on the following day. A small, round wound was found one and a half inches above and one inch behind the tip of the right mastoid process. The patient declared that the bullet had been extracted from the collar of his coat but an exit wound was not discovered. There was no œdema of the scalp. The pupils were small but reacted well and no abnormality was found within the territory of the cranial nerves.

Motion: The patient could move his head. Feeble adduction at the shoulders was the only voluntary movement possible in the upper extremities. He was able to flex and extend his right lower extremity at hip and knee, dorsiflex, extend and invert his right foot and move his right great toe feebly. He was just able to move his left hip and knee but no other voluntary movement could be performed in that limb. In respiration his chest moved as a whole; it was uncertain whether the diaphragm contracted or not. The abdominal muscles were paralysed.

Tone: The shoulder muscles were slightly rigid, especially the pectorals in which the rigidity was constant; the muscles of the arm, forearm, and fingers were flaccid. The abdominal muscles were in a state of tonic contraction and the lower extremities were rigid, the left more than the right.

Reflexes: Arm-jerks were obtained, left brisker than right; the abdominal

reflexes were absent on both sides; the knee and ankle jerks were increased, and ankle clonus was evoked on the left side; the plantar reflexes were of the extensor type.

Sensation: There was complete loss of sensibility to pin-prick in the right upper extremity, but pricks were appreciated in the left arm below the level of the fourth cervical root distribution. On the trunk the area of complete cutaneous analgesia extended in front on the right side to within an inch of the nipple, and on the left side to the level of the top of the xiphoid. The loss to pin-prick was incomplete over the upper part of the chest and the neck up to and including the area of skin supplied by the third cervical roots. Appreciation of cotton wool was lost below the third cervical root area. The patient was unable to recognize changes in passive position of his left upper and ower extremities, but he responded well when his right lower and occasionally when his right upper extremities were tested. Vibrations of a large tuning-fork (C_{128}) were not appreciated on the upper extremities or below the level of the clavicles except on the right lower limb.

Condition on February 12, 1915 (five days after the injury): The patient complained of a tingling sensation when his left arm was touched, but no other change in his sensory state had occurred. The following reflex reaction was evoked: Stimulation of the skin on the inner aspect of the elbow by means of three or four pricks evoked a strong involuntary movement of the upper extremity consisting of internal rotation and adduction at the shoulder. Internal rotation was the strongest component of the movement. The reaction was obtained equally well in each upper limb but was strictly unilateral. The movement, which the patient was unable to initiate by voluntary effort, started unexpectedly after the stimulus was applied, developed rapidly, quickly subsided, and was very strong. The response was confined to the shoulder-joint, the pectorals and subscapularis appearing to be the chief if not the only muscles engaged. The receptive field comprised the whole inner aspect of the arm from the internal epicondyle to the axilla, and the reaction could be easily evoked by pricking or pinching the skin or by stroking firmly along the inner margin of the biceps.

On February 16 (nine days after the injury) the patient was evacuated to England and it was noted that voluntary power over the lower extremities had improved before his departure.

For the subsequent history up to the time when he first came under our observation we have to depend on the patient's account only. From the first he had never experienced the feeling that he was dissociated from his body and limbs, but when he shut his eyes he thought that his arms were flexed at the elbow, whereas in reality they were continually in tonic extension. His trunk and lower extremities seemed to him to be extended. About two months after he was wounded he was able to extend his right hand feebly at the wrist and to move his right thumb. A little later voluntary flexion of his right elbow became possible and within another fortnight movements began to return in his left upper limb in the same order as those on the right side. There was also some recovery of sensibility in his upper extremities.

He was able voluntarily to empty his bladder five weeks after the injury and about a week earlier could tell when his bladder and rectum were full. Within twelve months voluntary micturition was almost perfect and the rectal contents could be expelled, although he has continued to require aperients.

Erections of the penis occurred fairly frequently, but emission of semen had not been noticed since he was wounded.

Summary of his condition from August 1, 1918 (*forty-two months after the injury*) *when he first came under our observation* :—

He is a well-educated man and an exceptionally good witness. His mental capacity is above the average and his critical powers are excellent. Speech and articulation have never been impaired; there is no history of fits or convulsions and he rarely suffers from headaches.

The left pupil and palpebral fissure are smaller than those of the right side. The pupils react well to light and accommodation, but the dilatation to shade of the left pupil especially is slight. The cilio-spinal reflex is present on the left and absent on the right side. Ocular movements are full and there is no strabismus, nystagmus or diplopia. He wears glasses to correct a slight refractive error in both eyes (myopia and astigmatism) but his visual fields are full and optic discs and fundi normal. There is no evidence of abnormal function within the territory of the other cranial nerves.

Motion: Voluntary movements.—Voluntary movements of the head and neck are excellent and he can elevate his shoulders well. His right upper extremity is less paralysed than his left and al gross movements are fairly well performed. At the shoulder-joints adduction of the arm is a powerful movement and is much stronger than abduction. On the right side he is able to abduct his arm to an angle of about 90°, but on the left side the range of the movement is less. He finds it easier to move his arm backwards than forwards and internal rotation is performed much better than external rotation.

The movements at the elbow-joints are full in range; extension is distinctly stronger than flexion. He is able to pronate his forearms completely, but supination is a weak movement; on the right side the forearm can be rotated through an arc of about 130°, while on the left side it cannot be brought farther than the position midway between pronation and supination.

Extension and flexion of the hands and fingers are fairly well performed and flexion of the fingers is stronger than extension. Two years ago he was unable to execute any movement of an individual finger without bringing all the other fingers of the hand into action. This was particularly so on the left side. But as time went on isolated movements of the fingers and thumb of the right hand became more possible, although innervation still tends to be widespread, especially if he expends more effort than usual. With his right hand he can now unbutton his coat and arrange his cards at "bridge" while he holds them between his left thumb and index finger; but he is unable yet to button up his clothes or to write legibly. He feeds himself with his right hand but is obliged to have his food cut up for him.

Extension of the trunk is stronger than flexion. Movements of the chest have improved considerably but the diaphragm is still weak.

The lower extremities are stronger than the upper and the movements of the right lower limb are better than those of the left. Extension at hip, knee and ankle are stronger than flexion, and adduction at hip than abduction. The weakest movement is dorsiflexion with eversion of the foot. Movement of the toes is comparatively free.

All voluntary movements, especially those of the upper extremities, are performed slowly and stiffly, the reaction time being prolonged in relaxation as well as contraction. In attempting to grasp the observer's hand quickly it takes him a few seconds to bring his muscles into action and many more have passed before the maximum contraction of the flexors is attained. A similar delay is evident in the relaxation of his grasp. Co-ordination of muscles in the performance of voluntary movements is as good as could be expected in the presence of muscular rigidity.

Involuntary and synergic movements: An involuntary movement of the upper limbs not infrequently occurs in response to stimuli of widely different origin. A sudden emotional disturbance such as may be caused by the slamming of a door, a strong tickling sensation in his nose or irritation of his cornea by an eyelash evokes the following reaction which is often confined to his right upper limb but may involve both. The arms, rigidly extended, shoot out straight a little in front of his trunk and jerk forwards and backwards from the shoulders. The movement develops explosively, continues for a short time and subsides fairly quickly. The trunk and lower extremities apparently take no part in the reaction.

Rapid rhythmic flexion-extension movements of the thumbs at the metacarpo-phalangeal joints are of common occurrence, but we have been unable to trace the stimuli which excite them.

Flexor jerks of the right lower extremity sometimes disturb him in bed, but they have never been frequent. If he stretches out his lower limbs before getting out of bed in the morning, his right leg often becomes rigid in extension and shakes violently for a few seconds.

Vigorous voluntary muscular contractions in one limb are usually accompanied by movements in the others. For example, when the patient attempts to flex his right knee against resistance, his left upper limb may abduct slightly at the shoulder, flex at the elbow, extend at the wrist and metacarpo-phalangeal joints and flex at the interphalangeal joints, while the fingers and thumb spread out. In his right upper extremity there may be adduction, internal rotation, and backward movement at the shoulder, extension at elbow, pronation of the forearm, slight flexion of the hand, extension of the fingers, and adduction of the thumb. In his left lower extremity strong extension of the whole limb with inversion of the foot occurs. Similarly a voluntary attempt to extend the right upper limb at the elbow evokes flexion of the crossed upper limb and bilateral extension of the lower extremities. These associated movements will be considered in greater detail in the next chapter.

Tone: The attitude assumed by the patient in standing is the following (fig. 1). His lower extremities are fully extended at the knees with the feet about eight or ten inches apart. He stoops slightly forward with his shoulders

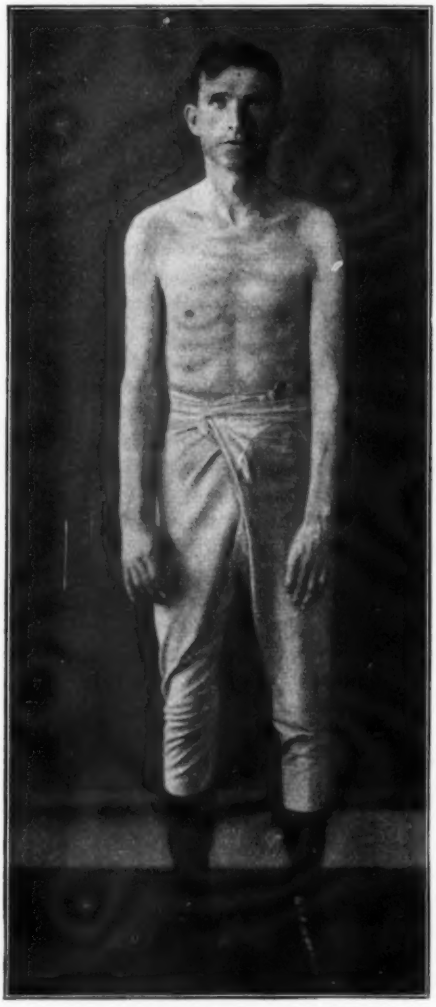

FIG. 1.—Photograph of No. 1 to show his general attitude in the erect position.

drooping, his arms a little in front of his trunk adducted and rotated inwards at the shoulders, rigidly extended at the elbows, the forearms pronated, the hands slightly flexed at the wrists, the fingers extended at the metacarpo-phalangeal

joints and flexed at the interphalangeal joints with hyperextension of the last phalanx of the thumb.

There is excessive rigidity of certain muscle-groups, especially in the upper extremities. The adductors of the arm, and in particular those of the anterior axillary wall, the pronators of the forearm and the flexors of the wrist and fingers, stand out prominently in tonic contraction and feel hard and tense when palpated. When muscle tonus is tested by the resistance offered to passive movement, it is found to be greatly increased in all the large muscles of the upper limbs, but in some more than others. The muscles with greatest tone are the adductors of the shoulders, triceps, the pronators, the flexors of the wrist and fingers, and the adductor-opponens group of the thumb. All the muscles of the trunk are hypertonic, but especially the erectores spinæ and the recti abdominales. The lower are less rigid than the upper limbs, but tonus is definitely increased, more so in the extensors than in the flexors. The left upper and lower extremities are more spastic than those of the right side.

Reflexes: In the upper extremities all the tendon jerks are increased. Any muscle when put on the stretch gives a brisk contraction in response to a tap on its tendon.

One of the interesting features in this case is a reflex reaction evoked by stimuli applied to the inner aspect of the upper extremity. Sensibility to pin-prick is defective along the inner aspect of the limb, yet when the skin in this region is firmly pinched the following response occurs explosively after a latent period of a few seconds. The arm suddenly shoots backward with extreme inward rotation, so that the palm of the hand faces almost directly outwards. There is adduction and internal rotation at the shoulder, extension at the elbow, pronation of the forearm, slight flexion at the wrist, and over-extension and adduction of the fingers. The fingers are so placed that they form a cone—the second finger overlapping the first and third, and the fourth underneath the third. The thumb is extended at the interphalangeal joint and adducted into the palm. The motor focus of the response is in the muscles of the shoulder-joint. When the reflex is excited while the patient is standing, the response is seen to involve the trunk and all four limbs, although it is most vigorous in the stimulated upper extremity. With extension, adduction, and inward rotation there is a backward thrust of the limb, the shoulder is pulled backwards, the upper part of the trunk rotates as a whole towards the stimulated side, and the patient is bent slightly forwards by the action of his abdominal muscles. The other upper limb becomes adducted at the shoulder and flexed at the elbow, but the movement is slight. The lower extremities remain extended at the knees and ankles.

The reaction outlasts the duration of a brief stimulus, so that the attitude provoked is maintained for many seconds and then gradually subsides, giving place to the original resting posture of the whole body.

One character common to all effective stimuli is the nocuous quality with which they are endowed. Pinching or scratching the palm of the hand, and the skin on the inner aspect of the limb and chest from the second to the

sixth costal cartilage are capable of evoking the response from either upper limb.

Reflexes are not obtained from stimulation of the upper or lower part of the abdominal wall, but can be elicited in the middle segment.

Cremasteric and bulbo-cavernosus reflexes are easily evoked on both sides.

Stimulation of the sole of the foot is followed by a typical flexion reflex consisting of flexion at hip and knee, dorsiflexion of the foot and upward movement of the toes. With a moderately strong reaction there is usually extension of the opposite lower extremity. The muscles of the abdominal wall do not always take part in the reaction. The receptive field for the flexion reflex extends from the plantar surface of the foot, where the threshold value of the stimulus is lowest, almost to the knee.

In response to stimulation of the skin or deep tissues of the thigh, extension at hip, knee and ankle and downward movement of the toes is usually obtained. If, however, the thigh is grasped suddenly and firmly, general flexion of the limb may result.

The knee-jerk on both sides is very brisk and with it there occurs adduction of the opposite thigh without flexion or extension of the hip.

The ankle-jerks are much increased and well-sustained ankle clonus can be evoked.

Sensation: The patient has been remarkably free from pain since his injury occurred. The only abnormal sensations of which he complains are numbness of the fingers, as if they were slightly swollen and cold, and "a feeling of tightness" in the muscles in front of his thighs, when he is bilious or out of sorts in any way.

Pin-pricks are appreciated as such, but with a raised threshold, over the post-axial aspect of the left upper extremity and hand, the pre-axial aspect of the right forearm and arm, and the shoulder and neck, below the level of the third cervical root distribution. On the right side of the trunk from the second rib to the groin pin-pricks are appreciated as points but fail to evoke definite pain. On the right lower extremity they give rise to a sensation of tickling only. Warmth is appreciated though with diminished acuity on the left side below the level of the third cervical root distribution. The defect is greatest on the lower extremity. Within the corresponding area on the right side the loss of sensibility to warmth is greater. Warmth is usually recognized correctly when the test objects are placed in contact with the upper limb and the chest, but below the costal margin the loss is complete. The loss of sensibility to cold is slight on the left side, but on the right side it is gross on the post-axial aspect of the upper limb and on the chest from the third to the seventh rib, below which level it is complete.

Tactile sensibility is not gravely disturbed except on the right side over the upper limb, the trunk below the second rib and the lower extremity. Complete loss to contact with cotton wool on hairless parts is limited to the right leg below the knee.

Vibrations of a large tuning-fork (C_{128}) are not appreciated on the trunk and

lower extremities below the first rib. On the upper extremities vibrations are appreciated only when the fork is vibrating well, but the defect diminishes towards the shoulder.

There is gross loss of recognition of passive position in all four limbs and the patient is entirely unable to recognize the shape, size and form of common objects placed in his hands. He has completely lost the power to estimate the differences in weights placed on his supported hands; tactile localization is disturbed less than the other spacial aspects of sensibility.

Vasomotor and nutritional changes: His skin is normal in colour and elasticity. There are no sores. In warm weather he sweats mainly from his head and neck and in these regions the hyperidrosis may be excessive.

Bladder, rectum and sexual functions: Control over the bladder is perfect. He is chronically constipated and requires to take aperients daily. Erections are frequent but there has been no emission of semen since the date of his injury.

Chapter II.—Analysis of the Reflex Phenomena in No. 1.

Posture: The attitude assumed by the body at rest is largely determined by the distribution of muscular tone. In the case of No. 1 the distribution of muscular tone was such that the posture of the trunk and limbs in the erect position or while recumbent was almost stereotyped; it varied within narrow limits only. In resting or sleeping he lay on his back with his head and shoulders slightly raised by pillows, his upper limbs lying by the side of his trunk in a position of general extension. The limbs were rotated inwards and adducted at the shoulders, extended at the elbows, with forearms pronated and wrists slightly flexed. His fingers were extended at the proximal joints and somewhat flexed at the interphalangeal joints; the first, second and third fingers were pressed together, while the fourth finger was a little abducted. The thumb, which was in apposition to the palmar surface of the index finger, was extended at the metacarpo-phalangeal joint and over-extended at the distal joint. His lower extremities usually lay extended on the bed, a little separated from one another, rotated outwards with the feet pointing.

The outstanding features of the general posture were the slight flexion of the neck and trunk from elevation of the head and shoulders, extension of all four limbs in line with the body and the strong inward rotation and pronation of the upper extremities. He was comfortable in this position for a long time and rarely moved much in his sleep. The extremities on the left side were more rigid than those on the

right and the upper more than the lower, so that if he desired to alter his position he did so by flexing his right lower limb and pushing with his foot against the bed, helping himself to some extent with his right arm.

But remarkable as was the patient's attitude when recumbent, the posture he assumed in standing was even more striking. The general features were the same in both positions, but the rigidity when erect gave him an almost statue-like appearance. This impression is illustrated by fig. 1.

The patient stood with his feet a little apart and his lower extremities fully extended at the knees; his trunk was slightly flexed with his head thrown back so that he could look directly forward. When he walked, however, he kept his eyes fixed on the ground a little in front of his toes. The photograph shows the strong tonic contraction of the upper fibres of trapezii, although the shoulders are drawn forward by the equally powerful contraction of pectorals. His upper limbs are rigidly extended and are adducted so that his hands lie in front of his thighs. They are turned inwards at the shoulders with the forearms pronated and the hands deviated somewhat radially at the wrists. The position of the fingers with extension at the metacarpo-phalangeal joints and flexion at the distal joints is well illustrated in the photograph. The thumbs are seen to be extended and pressed against the palmar surface of the first fingers.

Tone: The striking appearance of immobility of the patient when recumbent or erect was due to the excessive development of tone in his muscles, which, however, were not all equally loaded. The preponderance of tone in certain muscle groups was responsible for the constancy of the attitude of the body as a whole and of its component parts

The clinical tests at present employed in the investigation of the tonic state of muscles are crude and often untrustworthy, but in the case under consideration the distribution and certain other features of the rigidity were so distinct and constant that the following description can be accepted as fairly accurate.

All the muscles of the body from the shoulder-girdles downwards, with the exception of some of those of the fingers and toes to be detailed later, were in a state of increased tone. They felt firmer on palpation and presented more resistance to lateral displacement than the muscles of a healthy individual. The musculature of the extremities showed these features more on the left than on the right side, and that of the

upper extremities more than that of the lower. Detailed investigation revealed a greater increase of tone in some muscle-groups than in others. Thus in the upper extremities, and especially on the left side, the pectorals, triceps, pronators, and wrist-flexors were usually in a state of strong contraction. These muscles when palpated felt abnormally firm, and attempts to lengthen them passively were met by considerable resistance. Those of their antagonists, for example biceps and supinators which were readily accessible to direct examination, were found to be considerably less tonic and could be passively stretched with less difficulty.

The following tests brought out the difference in the tonic state of the different muscle-groups in another way. With the patient lying on his back the observer grasped one thigh and raised the limb from the bed. Instead of the knee flexing immediately as it would in a healthy individual the leg remained almost completely extended for a few seconds before the foot gradually sank towards the bed. Extension of the knee persisted for a longer time if the limb were lifted up quickly. It would seem that when the thigh was lifted the weight of the leg and foot excited an immediate reflex contraction of quadriceps cruris sufficiently strong and prolonged to keep the knee extended for some time against the influence of gravity.

This tonic rigidity of the lower limb in extension was increased and kept up for a longer time by evoking a flexion reflex in the opposite lower limb. For example, if the tonic reaction of the right quadriceps extensor was under observation and the point in the experiment was reached at which the leg began to fall towards the bed, complete extension could once more be obtained by scratching the sole of the left foot.

But, paradoxical though it may seem, it was possible to show that under certain conditions additional stimulation of the extensor reflex arc could temporarily decrease instead of augmenting the tonic extensor rigidity. The following experiment will illustrate the point: As before, the postural reaction in quadriceps extensor was evoked by suddenly raising the thigh from the bed, thus exciting the tonic reflex by mechanical tension. While the leg was being held in extension by contraction of the extensor of the knee, the patellar tendon was smartly tapped, thus supplying an additional brief stimulus of the same kind to the extensor reflex arc. This evoked a superadded contraction of the muscle in the form of a quick twitch, the foot first jerking upwards and then falling back again, but to a level lower than that at which it had been resting before the knee-jerk was excited. The jerk was

followed by three or four similar rapid movements of diminishing range before the leg finally came to rest. Although in the relaxation phase of each twitch the heel did not reach the bed, yet at the end of the reaction it was held considerably lower than at the beginning. In other words the knee instead of being in almost full extension was now somewhat flexed and complete flexion could be obtained as the end-effect of a series of knee-jerks.

Analogous reactions were obtained in triceps when the upper arm was passively supported in abduction and internal rotation, so that the weight of the forearm and hand pulled on triceps.

A different result, however, was obtained on investigation of the tonic reaction to gravity in the flexors of the elbow and knee. For example, in the case of the lower extremity with the patient lying on his face the leg was bent at the knee to almost a right angle, and then quickly released. Instead of being "held" by tonic contraction of the hamstrings in the position in which it had been passively placed it began almost immediately to extend and fell more rapidly than in the experiment with quadriceps extensor. So also with the flexors of the elbow, the tonic reaction excited by sudden tension on the muscles exerted by the weight of the limb was relatively much feebler than in triceps.

The unequal distribution of tonus was demonstrated in another way. As already mentioned, the voluntary movements which were most powerful were those in which the more hypertonic muscles were brought into action as prime movers. In the upper limbs these movements were adduction and internal rotation at the shoulder, extension at the elbow, pronation of the forearm and flexion of the hand and fingers; in the trunk extension and in the lower extremities extension and adduction at the hip, extension at the knee and downward movement of the foot. The muscles which determined these movements are exactly those which by their action determined the resting posture of the body. But not only were the movements just enumerated the most powerful, they were also performed most rapidly to command. Thus when on command the patient attempted quickly to abduct his arm, an interval of several seconds elapsed before the desired movement was begun, despite obvious efforts on the part of the patient to carry out the action promptly. The reaction time was long and the movement once started was slow and deliberate. The slowness of the response was all the more evident when for the performance of the movement it was first necessary to relax antagonists

which were in voluntary contraction. For instance, a longer interval elapsed between the giving of the command and the commencement of the movement of extension of the fingers when to carry out the action the patient had first to relax his grasp of the observer's hand, than when the fingers were primarily in the position of rest in slight flexion.

In contrast to the deliberate character of these movements was the greater rapidity with which those muscles that were overloaded with tone were voluntarily contracted. Thus the patient when asked to flex his fingers or adduct his arm carried out the movement comparatively quickly and with considerably less effort.

An outstanding feature of the rigid muscles was the way in which they reacted to passive lengthening. When the observer flexed the elbow, the patient meanwhile remaining entirely passive, and exerted a *steady* pressure throughout the movement, he became aware of an increasing resistance in triceps which developed fairly quickly to a maximum. This resistance was maintained at its height until flexion was almost complete and then rapidly declined. If the forearm was flexed through part of the arc only and was then released by the observer, extension at the elbow sometimes immediately followed. This reaction was due to the contraction of triceps which had been not only maintained, but considerably increased by the stimulus of passive lengthening of its fibres. Further observations, however, showed that the additional amount of tonicity caused by the mechanical tension on the muscle disappeared, and the new muscle-length was "accepted" provided that the tension stimuli were interrupted. Thus, when the elbow was suddenly flexed in short jerks by the observer the resistance offered by triceps was felt at the beginning of each movement to increase more rapidly than when flexion was carried out slowly. If an interval was allowed to elapse between each jerk, it could be readily ascertained by palpation of triceps, and by "sensing" the tension of the muscle by slight movement at the elbow, that the accretion of tone due to the sudden stretching of the fibres quickly disappeared. In other words, the muscle had assumed a new length and was being "held" by a tonic contraction of approximately the same strength as before it was disturbed by the passive movement. The muscle recovered its original tonic contraction for each new length. This phenomenon was evident over a wide range of different lengths of triceps, but was minimal or absent when the limb was in positions approximating complete flexion or extension at the elbow. The same phenomena

were evident in the majority of the other muscles belonging to the rigid group. Thus it was brought out by passive flexion of the knee and extension of the wrist. But in those muscles of this group in which tonicity was most developed, namely, the pectorals and pronators, the resistance encountered on steady passive stretching increased very rapidly, was maintained at its height throughout the process of displacement and did not appreciably diminish during the time that the limb-segment was being held in the new position by the observer. Further, unless the process of lengthening the muscles was carried out slowly the tension was apt to excite a powerful reflex movement of the limb as a whole, in which inward rotation and pronation with extension at the elbow formed outstanding components. With careful manipulation, however, the arm could be completely adducted, or the forearm completely supinated.

Examination of the group of muscles which exhibited a less degree of tonic contraction than those we have been studying showed that in them a lengthening reaction was inconstant and when present was in comparison poorly developed. When, for example, the arm was extended at the elbow by quick jerks, the resistance to the passive movement offered by the flexors was usually increased but to a less extent than with the triceps during passive flexion. Another point of difference was that the forearm had to be displaced through a larger arc before the muscular tension notably increased. The flexors of the elbow when passively lengthened appeared to become "gathered up" and to "set" in a new posture more slowly and less completely than the extensors.

It was possible to demonstrate that triceps and the extensors of the knee could accommodate themselves to new postures which involve shortening without appreciable diminution of tone. Thus, if the observer extended the elbow by moving the forearm with one hand while he grasped the triceps with the other, he was able to satisfy himself that, as the points of attachment of triceps were approximated, the muscle did not become flabby, but on the contrary remained firm to the touch. That is to say the tonic functions of triceps and quadriceps cruris were such that these muscles were maintained in approximately the same degree of contraction although the distance between their attachments was progressively diminished.

During a voluntary movement the muscles not concerned directly in bringing about the required displacement of the limb segment displayed certain changes. The antagonists relaxed and the contraction of the muscles concerned with the fixation of joints, necessary for the move-

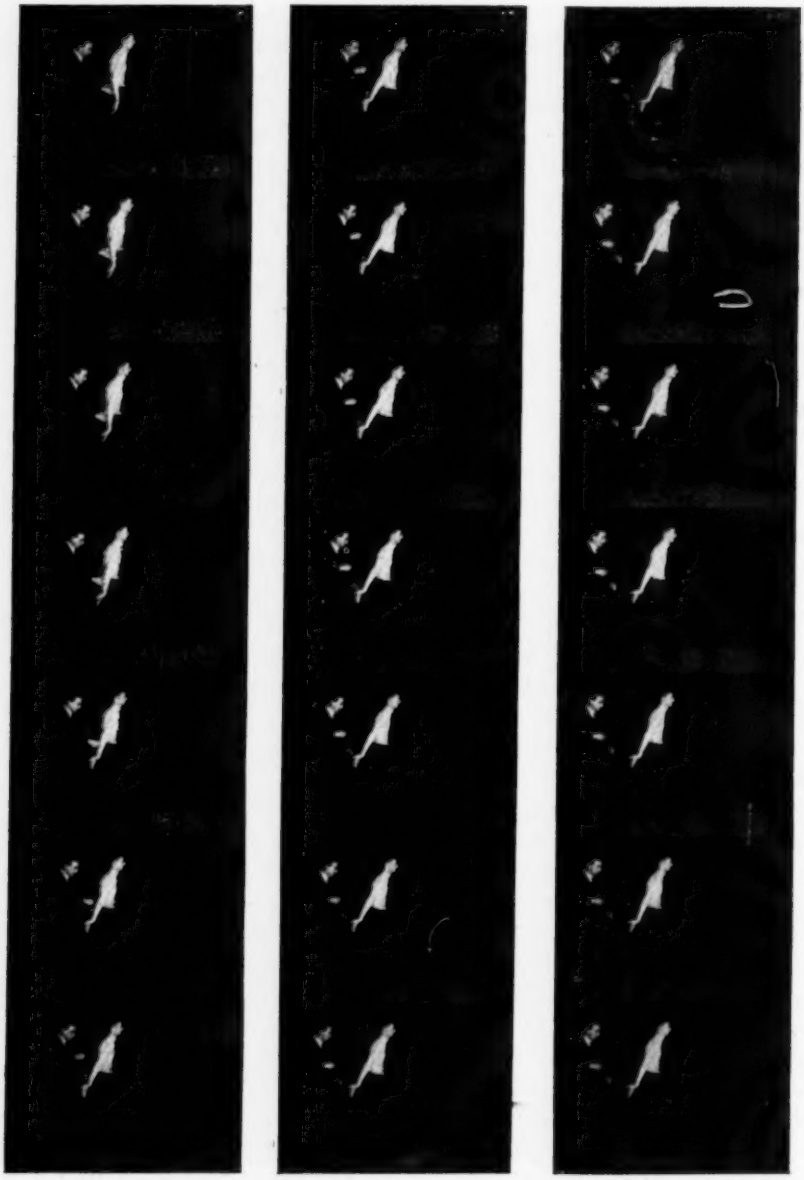

FIG. 2.—A series of cinematograph photographs showing the extension reflex reaction of the upper limb in No. 1 (side view).

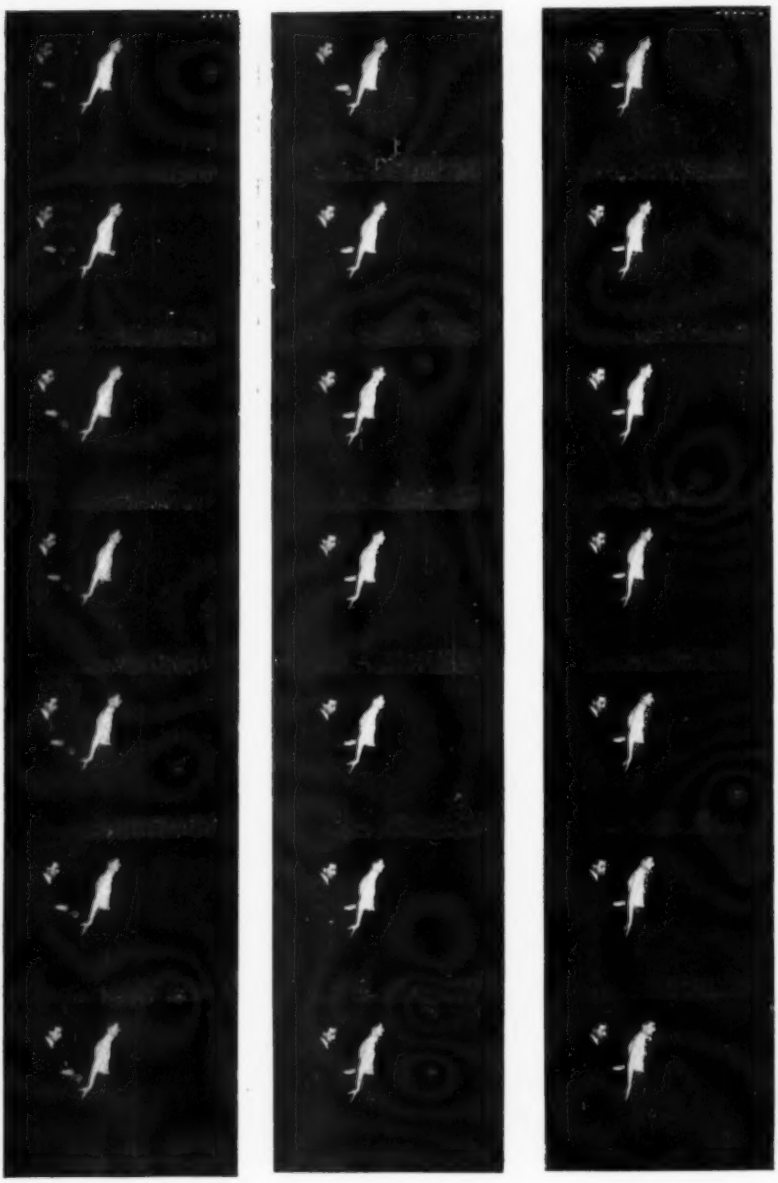

Fig. 2 continued.

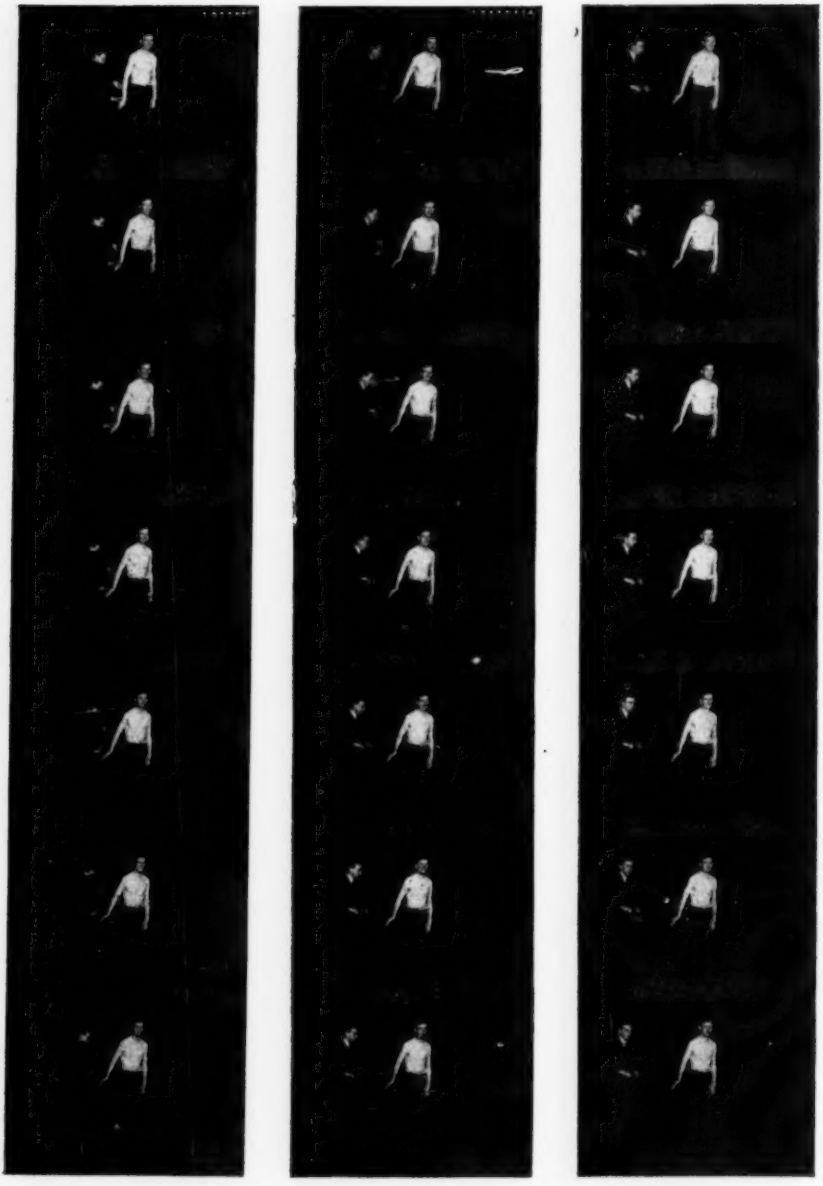

FIG. 3.—To show the extension reflex reaction of the upper limb in No. 1 (front view).

Fig. 3 continued.

ment, increased. If a strong voluntary effort was necessary to bring about the desired movement the tonic state of all the muscles of the limb might be altered. For example, when the patient was asked to squeeze an object firmly with his hand, all muscles playing on the wrist, elbow and shoulder-joints became more powerfully contracted; for the moment they were acting as fixators.

We have so far been dealing only with the tone of the large muscles of the trunk and extremities. The muscles of the toes, however, and certain of those which play on the finger joints have in this respect to be dealt with in a group by themselves. The toes and the fingers were freely movable in every direction and indeed the latter, especially the middle, ring and little fingers, could be hyperextended to an extreme degree without difficulty. The adductors of the thumb, however, were involved in the general rigidity and showed reactions to passive movement which were similar to those described for triceps, pronators and the other more hypertonic muscles.

Extension reflex of the upper limb. — Early in our investigations on No. 1 we discovered that an unusual and striking reflex reaction could be obtained in the upper extremity on either side in response to nocuous stimulation of the limb. With the patient lying in bed on his back the position of his upper limbs, it will be remembered, was one of adduction and inward rotation at the shoulder, extension at the elbow, pronation of the forearm and slight flexion at the wrist; the hands were not as a rule deviated to one or other side; the fingers were extended at the metacarpo-phalangeal joints and slightly flexed at the interphalangeal joints, while the thumb in extension was pressed against the palmar surface of the index finger.

On drawing the point of a pin or the tip of a finger-nail firmly along the ulnar aspect of the hand or forearm the following displacement of the limb occurred. There was strong adduction, internal rotation and retraction at the shoulder, extension at the elbow, extreme pronation of the forearm, increased flexion at the wrist, ulnar deviation of the hand, over-extension and adduction of the fingers and thumb. The fingers overlapped one another like the slates on a roof, the middle finger being posterior and lying over the index and ring fingers, while the thumb with extended phalanges was tucked into the palm and more or less in line with the middle finger.

The reaction could be observed more completely when the patient was standing and, in general, consisted of a vigorous movement by which the upper limb was adducted, extended and thrust backwards

with extreme inward rotation of the limb as a whole, so that the palm of the hand was directed backwards and outwards (figs. 2 and 3).

Although certain of the muscles involved could be seen and felt to contract almost immediately on application of the stimulus, usually an appreciable length of time elapsed before the movement took place. It was as if the impulses excited by the stimulus had to break through a strong resistance or had to reach a certain maximum intensity before they were capable of evoking the response in full. This supposition gains support from the fact that a brief harmful stimulus was as a rule ineffective; a prolonged stimulus was often required. For example, a single prick usually produced very little effect; the point of the pin had to be dragged across the skin and, although the pectorals contracted almost from the commencement of stimulation, the distance traversed by the pin often reached two or three inches before the limb was displaced. But once the point of sufficient intensity of stimulation was reached the reaction, although it could be graduated to some extent, tended often to occur explosively and the movement was sometimes so violent as almost to throw the patient off his feet.

Relaxation of the contracted muscles did not immediately follow the full displacement of the limb (figs. 4 and 5). The new position was maintained for several seconds and then the limb slowly resumed its original posture at rest.

Usually the reflex was experimentally excited with the upper extremity in the resting attitude of extension, adduction and pronation; but investigations showed that alteration of the initial position of the limb, for example flexion at elbow carried out either by the observer or by the voluntary effort of the patient, did not lead to any material change in the response.

A point of considerable interest is that the patient seemed to have little power to inhibit or control the reaction.

The muscles mainly concerned in bringing about the movement were those which were overloaded with tone and in consequence determined the attitude of the limb at rest. Incidentally attention may again be drawn to the fact that it was exactly those muscles which were rigid and which acted as prime-movers in the extension reflex that could voluntarily be brought into action most strongly and readily by the patient.

The condition of the antagonists in the reaction was noteworthy. Deltoid, biceps and supinator longus from their superficial position could readily be investigated by palpation. They were certainly actively

employed in the reflex when it was fully excited; they became firmer and could be transversely displaced less easily although their tonic contraction was less than that of the prime movers. Even by the rough methods of testing employed it was also possible to satisfy ourselves that the increase in tone of biceps was proportionate to the increased contraction of triceps. In other words, there was a definite relation between the state of the two opposing muscles. It must be clearly understood, however, that these statements hold good for the period of

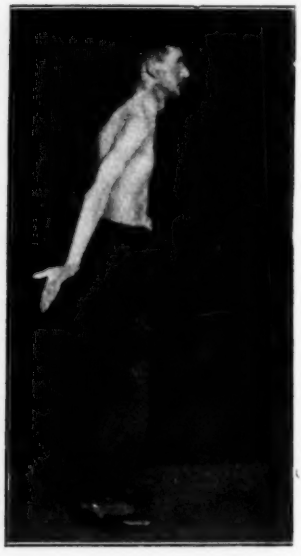

FIG. 4.—Photograph showing the attitude of the right upper limb at the height of the extension reflex reaction in No. 1 (side view).

FIG. 5.—Photograph showing the attitude of the right upper limb at the height of the extension reflex reaction in No. 1 (front view).

time only in which the new posture of the limb was being maintained. It was impossible by our methods to ascertain the tonic state of the antagonists whilst the arm was being displaced in one or other direction by the reflex.

The receptive field of the reflex was extensive, comprising almost the whole limb and invading the chest in front and behind. The part of the field in which the threshold value of stimulus was lowest was the skin covering the walls of the axilla, but the threshold was not greatly higher for the inner aspect of the arm, forearm and palm of hand.

Within this area a moderately firm scratch with a sharp object or a prolonged squeeze of the skin or deep tissues formed an appropriate stimulus. The reflex was readily excited by pinching the tendons of biceps, pectoralis major or latissimus dorsi or by attempting suddenly to rotate the arm outwards at the shoulder or to supinate the forearm.

A response was never obtained from stimulation of the dorsum of the hand or fingers but was occasionally elicited when the stimulus was applied to the skin on the outer aspect of the limb. The receptive field on the front of the chest covered an area which extended from about the third to the sixth ribs, and on the back roughly corresponded in size and position to that on the front. Above and below these levels we were never able to excite the reflex with the stimuli we employed.

The reaction varied within narrow limits with the situation of the stimulus. Thus when the reflex was excited from the palm, movements of the fingers occurred early in the experiment and before the general reaction was evoked. Most often the preliminary movement consisted of extension and abduction of the fingers and thumb, while in the general reaction the digits were strongly adducted. Pronation of the forearm was a prominent early movement when the site of the stimulus was on the ulnar aspect of the hand or forearm, and diminution of the pronated resting attitude, usually with abduction of the thumb, followed application of the stimulus to the thenar eminence. The outstanding early movements which resulted from stimulation of the inner aspect of the upper arm or the chest were adduction and internal rotation at the shoulder and extension at the elbow. Indeed, from whatever point within the receptive field the reflex was evoked the muscles of the anterior axillary wall could almost invariably be seen to contract early. It may be remembered that attention was drawn on p. 400 to the response obtained by Lieut.-Colonel Gordon Holmes five days after the injury. Nocuous stimulation of the skin on the inner aspect of the arm between the axilla and elbow evoked a strong involuntary movement of internal rotation and adduction at the shoulder; at this time no movement at the elbow was observed. He especially noted that inward rotation of the arm was the chief component of the movement and predominated over adduction. This movement has remained the minimal response for the whole period during which the patient has been under our observation.

Two noteworthy variations in the reaction were sometimes observed by us when the appropriate stimulus was of a special kind and applied in the neighbourhood of the shoulder. While investigating by passive

movements the tonic state of the internal rotators at the shoulder a reflex movement of the whole upper limb was often excited, especially if the observer suddenly stretched the muscles. At times the movement consisted of the adduction-pronation-extension response already described, but often the reaction was more complicated. What happened was briefly as follows. When the arm was rotated quickly outwards the resistance encountered steadily increased if the attempt to overcome it was persisted in. The suddenly applied tension stimulus had succeeded in evoking superadded contraction of the internal rotators, the strength of which increased proportionately to the strength of the stimulus. With internal rotation at the shoulder there occurred flexion at the elbow and wrist, flexion and abduction of the fingers and extension and abduction of the thumb. This posture was broken into from time to time and replaced for short periods by extension of the elbow, over-extension and adduction of the fingers and adduction of the thumb.

The second pronounced alteration in the extension reaction was sometimes observed when the reflex was evoked by pinching the muscles of the anterior or posterior axillary walls. It consisted of a rapid, rhythmic, flexion-extension movement at the wrist instead of the more usual maintained attitude of flexion. The reaction at the shoulder, elbow, finger and thumb joints did not, however, differ from that obtained by stimulation elsewhere.

There were no essential differences in the reflexes excited in the two upper extremities.

Motor phenomena accessory to the extension reflex of the upper limb.—As a rule the motor effects of the reflex spread beyond the stimulated upper limb and involved the trunk and other three limbs. The trunk and lower limb on the same side were affected as soon as or earlier than the other upper limb. The associated reaction consisted of (1) contraction of all the muscles of the trunk and of the three limbs—those of the shoulder and hip joints being first involved, and (2) movements which usually were slow and deliberate and were due to greater contraction of some muscles than of their antagonists. The most common associated displacements were slight adduction and external rotation at the shoulder, flexion at the elbow and slight extension at the wrist in the opposite upper limb; the trunk was usually rotated and became slightly flexed and the lower limbs remained extended at the knees and ankles. The serial cinematograph photographs on pp. 412 to 415 represent a general reaction of this kind.

Motor phenomena in association with the flexion reflex of the lower extremity.—Stimulation of the sole of the foot (pinching, pricking or scratching) evoked a typical flexion reflex of the lower extremity consisting of flexion at hip and knee, dorsiflexion at ankle and upward movement of toes. The plantar aspect of the foot formed the focus of the receptive field which, for stimuli of moderate intensity, reached as high as the knee. Stimulation of the thigh as a rule evoked extension at hip and knee and downward movement of the foot and toes, but if the stimulus was intense (firm squeezing of the thigh) flexion of the limb frequently formed the primary response. Contraction of all the limb-extensors was readily excited by sudden passive dorsiflexion of the foot.

The strength of the flexion reflex could be varied to some extent by grading the intensity of stimulus, and the motor effect in a mild reaction was usually confined to the stimulated lower extremity. But by intensifying the appropriate stimulus it was possible to obtain a more widespread reaction the extent of which could also be controlled more or less by regulating the strength of stimulus. In the order of spread of the reflex the opposite lower extremity was first affected, then the muscles of the abdominal wall and back and lastly the upper limbs.

In the associated reaction all the muscles acting on a joint were contracted, but usually some more strongly than others, so that movements resulted. Movements, however, were sometimes absent, especially in weak reactions, and when present were never excessive and developed slowly. The crossed lower limb remained extended at hip and knee and the foot and toes moved down. The upper limbs became rotated outwards at the shoulders, flexed at the elbows, extended at the wrists with slight supination of the forearms; the fingers became extended at the metacarpo-phalangeal joints and flexed at the interphalangeal joints while the thumbs were extended and abducted. The muscles of the proximal joints of the limbs were always affected earlier than those nearer the periphery of the limbs. If the stimulus was weak the upper and lower limbs almost immediately resumed their resting attitudes, but in a strong reaction there was a definite after-action so that the postures were maintained for a short time before the limbs slowly resumed their resting positions.

Motor phenomena in association with voluntary movement of one limb against resistance.—Voluntary efforts of the patient to move one of his limbs or a segment of it when strongly resisted by the observer as a rule produced alterations in the attitudes of some or all of the other

limbs. These adjustments were not constantly the same on different occasions for any one voluntary movement. As with the general reactions associated with the extension reflex of the upper limb and the flexion reflex of the lower limb, the essential feature of the response was increased contraction of antagonists as well as prime movers so that movements when present were slow.

With forced voluntary movements of flexion or extension of the right lower extremity the response in the left was almost invariable extension. If at the beginning of the experiment the patient was lying in bed with his lower limbs stretched out, the only segments of the left lower limb which moved were the foot and the toes and these were thrust downwards, but the flexors as well as the extensors of the knee and ankle were in strong contraction, the extensors predominating. In a strong reaction extension of the crossed limb was accompanied by internal rotation at the hip.

The response in the right lower extremity in association with forced voluntary flexion or extension movements of the left lower limb differed as a rule in certain particulars from the reaction which has just been described. The want of correspondence may have a definite relationship with difference in the development of resting tonus in the muscles of the two limbs; the left was more rigid in extension than the right. The main points of divergence lay in the reactions at hip and knee, for when the right was the crossed limb some degree of flexion was usually observed. It must be understood, however, that although slight flexion occurred in the early phase of the response the movement formed a minor part only of the whole reaction which comprised a tonic contraction of all the muscles of the limb.

The movements and postures in the upper limbs induced by the voluntary efforts of the patient to move one lower extremity against resistance were subject to greater variation than those we have just studied in the crossed lower extremity. The principal variation was in the direction of the movement at the elbow, but it did not seem to occur in one limb more than in the other; neither did it apparently depend on which lower limb the patient was attempting to move nor on the direction of the movement, whether flexion, extension, adduction or abduction. The reactions in the upper limbs were also feebler than the response in the crossed lower limb. Three main reflex-figures were observed in the upper limbs: (1) Internal rotation and adduction at shoulder with extension at elbow on the side of the lower extremity

which was being voluntarily displaced by the patient, and external rotation at shoulder and flexion at elbow in the upper limb on the opposite side. (2) Bilateral adduction at shoulder, extension at elbow and wrist with extension and abduction of the fingers at the metacarpo-phalangeal joints, and flexion at interphalangeal joints and abduction of thumb (the so-called "claw" position of the hand). (3) The same posture in both upper limbs as last described except that the elbows were slightly bent.

With forced voluntary movements of the upper limbs the lower extremities invariably remained in extension, although the tonic contraction of all their muscles was increased. The response was stronger in the lower limb on the same side and here extension was often accompanied by adduction and inward rotation.

In contra-distinction to what obtained in the lower limbs the direction of voluntary displacement of the forearm usually altered the posture of the opposite upper limb at the elbow. Thus forced extension of the elbow on one side was accompanied by flexion of the elbow on the other side; the movements on the two sides were usually of opposite direction.

In each general response all the muscles of the trunk were brought into action.

As has already been pointed out, all the associated reactions which have been described in this section were made up of two components: (1) the maintenance of attitudes or postures due to the tonic activity of prime movers and antagonists; and (2) displacement of segments of the three limbs brought about by predominant contraction of certain muscle-groups.

Involuntary movements of the upper limbs excited by stimulation of parts outside the paralysed regions of the body and by changes in the patient's mental state.—Sometimes as the patient lay quietly in bed or was sitting up in a chair his arms suddenly shot out in front of him, extended at elbow, wrist and finger joints, and flapped up and down from the shoulders for several seconds. The occurrence of these involuntary movements could not be traced to irritation of the limbs or trunk or to voluntary effort on the part of the patient, and we were at a loss how to explain them. The patient, however, soon noticed that there was a direct relationship between them and the development of a sensation of tickling in his nose, or a sensation of irritation of the surface of his eyeball, such as is caused by an inverted hair of the lower lid. He was particularly subject to disagreeable sensations in his

nose so that the rhythmic movements of his upper limbs occurred frequently.

These involuntary movements were, however, evoked not only by stimulation of the nostrils and eyeballs, but also frequently followed a sudden emotional disturbance. For example, if the patient was suddenly startled by the slamming of a door or by some other cause his upper limbs were immediately thrust out and began to move up and down from the shoulder. The movements never continued for long at a time but he had little power to control them.

Chapter III.—An Example of Spastic Hemiplegia.

Case No. 5.—A. C., aged 41, house painter. At the age of 18 years he developed a sore on his penis, which was untreated. He married at the age of 21 and his wife has borne him three healthy children. There is no history of miscarriages. Nine years ago the patient was treated in the Venereal Department of Guy's Hospital for an ulcer on his tongue. On Easter Sunday, 1919, while out walking, he was suddenly seized with severe pain in his right wrist and shin. It passed off in a few seconds and he was able to walk home. When he awoke the next morning he found that the right half of his body was paralysed and he was unable to speak.

He received a course of anti-syphilitic treatment in Guy's Hospital in May and June, and he recovered sufficiently to be able to walk with the help of a stick. In November, 1919, he came under our observation and presented the following features.

He was a fairly intelligent man with a good memory. Speech was defective, but he could answer questions and express his thoughts in speech if given time. In the evenings he complained of a sensation of freezing in the left side of his head, but he was free from headaches. He did not suffer from fits. His visual fields were full and his fundi normal. He showed weakness of voluntary movement on the right side of his face; his tongue when protruded deviated to the right, and his jaw and palate movements were weak on the right side, but associated facial movements were stronger on the right than on the left side. Otherwise there was no defect in the functions of the cranial nerves.

All voluntary movements of his right upper extremity were weak, those of the distal segments being most affected. Abduction of the shoulder, flexion of the elbow, pronation and supination of the forearm, extension of the wrist and fingers were particularly weak and voluntary movements of the individual fingers were impossible.

His right lower extremity was stronger than the right upper limb; voluntary extension and adduction of the hip, extension of the knee and downward movement of the foot were powerful and considerably stronger than the other movements of the limb.

The voluntary movements of the left side of his face and of the left upper and lower extremities were unaffected.

The tendon jerks of the upper and lower limbs on the right side were brisker than those on the left. The right abdominal reflexes were diminished in the upper and middle segments and absent in the lower. Cremasteric reflexes were obtained on both sides. The right plantar reflex consisted of upward movement of the toes and foot and flexion of the knee and hip. This reaction was obtained by pinching any part of the leg below the knee as well as by scratching or squeezing the foot. The threshold of stimulus was lowest in the sole of the foot. No associated movement in the opposite lower limb was observed, but flexion of the left knee was evoked on several occasions by squeezing the muscles of the right thigh. The minimal response to plantar stimulation on the left side was downward movement of the toes and contraction of tibialis anticus ; a stronger stimulus evoked, in addition, upward movement of the foot and flexion of the knee and hip, and in the opposite lower limb contraction of the extensors of the hip and knee and downward movement of the foot and toes.

Sensibility to light touch, pin-prick, thermal stimuli and changes in passive position were not diminished, and recognition of size, shape and form was as good with the right as with the left hand. The threshold for pain evoked by deep pressure (algometer) was lower on the affected than on the normal hand. Vibrations of a tuning-fork (C_{128}) were appreciated better on the left than the right side.

At first he had slight difficulty in starting micturition, but this disappeared later. Aperients were often required to overcome constipation.

Vasomotor, sudorific and nutritional changes were not observed.

Posture.—The right upper limb was slightly abducted and internally rotated at the shoulder, flexed 45° at the elbow, three-quarters pronated, hand in line with forearm, thumb extended at metacarpo-phalangeal joint and slightly flexed at interphalangeal joint, and fingers three-quarters flexed at all joints.

The right lower limb was slightly rotated outwards at hip and extended at all joints with the foot turned slightly inwards.

When the patient was standing a slight droop of the right shoulder was noticeable in spite of the fact that trapezius on that side could be seen to stand out in strong contraction. At the same time the attitude adopted by the trunk was one of slight flexion.

Tone in the right upper limb was generally increased but distinctly more so in the adductors of the shoulder, the flexors of the elbow, the pronators of the forearm, and the flexors of the wrist and fingers than in their respective antagonists. There was less resistance to passive flexion of the fingers than to movements at any other joint. In the lower limb tone was relatively greater in the extensors than the flexors, in the adductors of the thigh than in the abductors, and in the invertors of the foot than the evertors. In the trunk the extensors were on the whole more loaded with tone than the flexors.

The phenomena associated with increased tone, which we have already described in relation to our quadriplegic patient in the last chapter, were mani-

fest in the case of hemiplegia under consideration as well as in others under our observation. There is nothing to add to our description on p. 408 of the shortening and lengthening reactions, and the responses to the tests employed had, in general, the same features.

Flexion reflex of upper limb.—When the palm of the hand was scratched or pinched, the following movements were observed: Slight flexion of the fingers and wrist, more powerful flexion of the elbow, usually pronation of the forearm, abduction and external rotation and elevation of the shoulder. The threshold of stimulus was lowest in the palm of the hand, but a response could be easily obtained by scratching or pinching the skin on the inner aspect of the arm and forearm or that on the chest, behind and in front, corresponding roughly to the distribution of the third and fourth thoracic segments.

Reciprocal relaxation of the adductors (pectorals) accompanied contraction of the abductors and external rotators of the shoulder, including latissimus dorsi. With contraction of biceps there was usually relaxation of triceps.

The response varied slightly in detail with the locality of the stimulus. Thus scratching the radial side of the palm evoked abduction with extension of the thumb and some supination of the forearm. When the stimulus was applied to the inner aspect of the forearm near the elbow flexion of that joint was exaggerated, and the same observation applied to external rotation and abduction of the shoulder when the skin of the anterior wall of the axilla was pinched. Elevation and retraction of the shoulder were often observed following stimulation of any part of the reflexogenous zone.

The influence of the locality and duration of the stimulus were well shown by the response evoked by drawing the point of a pin from the elbow to the palm of the hand along the inner aspect of the limb. The response began with abduction, retraction, and elevation of the shoulder and flexion of the elbow; as the stimulus approached the wrist, this joint became flexed, and when the palm was reached a movement of flexion of the fingers appeared. The response continued so long as the limb was being stimulated; with removal of the stimulus the muscles gradually relaxed.

Single pricks, when effective, evoked a momentary quick response.

This reflex movement was more deliberate and less explosive in character than the extension reaction of the upper limb in quadriplegia and, as sometimes seen, in hemiplegia (No. 8).

Motor phenomena in association with the flexion reflex of the right upper limb.—With strong reflex flexion of the right upper limb there occurred contraction of deltoid, pectorals, triceps, and possibly biceps of the left side, although no displacement of the left upper limb actually occurred. All the muscles of the trunk on both sides were contracted. In the lower limbs all the muscles acting on hips, knees, and ankles became strongly contracted, those on the right more than those on the left side. Both remained extended.

Motor phenomena in association with the flexion reflex of the right lower limb.—All the muscles of the left lower limb became contracted, but the limb remained firmly extended. The trunk muscles were called into action on both

sides in front and behind. In the right upper limb there was strong contraction of the abductors and adductors of the shoulder; of the flexors and extensors of the elbow with a slow and incomplete movement of extension; of the flexors and extensors at the wrist with extension at this joint; and of the flexors of the fingers so that the hand became clenched. Sometimes rapid movements of extension alternating with flexion of the wrist were observed. In the left upper limb both adductors and abductors of the shoulder and flexors and extensors of the elbow were brought into action, but no displacement of the limb could be detected.

Motor phenomena in association with voluntary movements of one limb.— With powerful voluntary flexion or extension of the left elbow against resistance a strong reaction occurred in the right upper limb, consisting of increased contraction of all the muscles, but of some more than others, with the result that the following displacements occurred: Adduction and internal rotation of the shoulder, incomplete extension of the elbow, strong extension of the wrist with complete flexion of the fingers. Alternating flexion and extension movements at the wrist were sometimes noticed. All the muscles of the trunk were brought into action, the extensors more strongly than the flexors. There was bilateral extension of both lower limbs with increased contraction of all the muscles. The reaction was more powerful on the right side, where it was accompanied by inversion of the foot.

When the right elbow, that of the paralysed side, was voluntarily flexed against resistance, the reaction in the lower limbs was the same as that just described. The response in the crossed (left) upper limb was feeble in comparison with that obtained in the right upper limb, when the left (non-paralysed) upper limb was the seat of voluntary effort. It was confined to the proximal joints, and included slight flexion instead of extension of the elbow.

With voluntary flexion or extension of the left knee against resistance there was usually extension of the trunk and of the other three limbs with general increase of muscular contraction. Sometimes with voluntary flexion of the left knee there was flexion of the right knee and of the right elbow.

In association with voluntary extension of the right (paralysed) knee, there occurred extension of the trunk and of the other three limbs, the reaction being stronger in the right than in the left upper limb.

Speaking generally, the reaction in the remote limbs was stronger when the voluntary effort was being made in one of the lower limbs than when one of the upper limbs was the seat of the voluntary action.

It was also noticeable that the reaction of the fellow limb was more strongly developed when the voluntary effort was directed to a movement of a lower limb than to one of an upper limb.

The outstanding feature of all these associated reactions was the tonic contraction of antagonistic pairs of muscles involving in the first place the musculature of the trunk and proximal segments of the limbs, and spreading, as the voluntary effort increased, to more distant segments.

The contraction of some muscles exceeded that of their antagonists, with the result that segments of limbs were displaced. This displacement was slow, deliberate, and rarely excessive; its direction at any given joint did not usually alter throughout a reaction. This rule had some exceptions, as for example in the alternating movements sometimes observed at the wrist.

Finally, the reactions were less easily evoked in the healthy than in the paralysed limbs, and when present were usually confined to the proximal muscles and unassociated with displacement. Nevertheless, it was clear that though differing in intensity, the reaction on each side was of the same order.

Chapter IV.—Reflex Movements of the Upper and Lower Limbs in Quadriplegia and Hemiplegia.

(*A*) *Appropriate Stimuli.*

One of the reasons that led Sherrington to apply the term nociceptive to certain reflex reactions, such as the flexion reflex, in decerebrate and spinal animal preparations, was that they are constantly and most easily excited by potentially painful and harmful stimuli. Any form of nocuous stimulus is suitable, but the degree of noxa required varies under different conditions; sometimes protective reactions may be evoked by stimuli which in the normal individual would produce sensations of mild discomfort only. Thus after complete transection of the spinal cord in man vigorous flexion of the lower extremity may follow that form of light stimulation of the sole of the foot which is usually associated with a sensation of tickling ([6] p. 305). But as a rule such stimuli are effective when applied within the focus of the receptive field only and after a period of excitation of several seconds. In addition the isolated portion of the spinal cord must have reached a high degree of reflex excitability, and have become, as it were, habituated to flexor reactions by frequently recurring involuntary flexor spasms.

Under ordinary clinical conditions, however, it is comparatively rare to find an injured spinal cord that is so exquisitely responsive to stimuli; more often appropriate stimuli for the flexion reflex of the lower limb are of the painful order.

To elicit nociceptive reactions of the upper or lower limbs in our cases of hemiplegia and quadriplegia potentially painful stimuli were usually necessary. Pricking or scratching the skin with a sharp pin or pinching the tissues were usually employed. Hot or cold objects

were also effective provided their temperature was high or low enough to be capable of exciting a sensation of pain when the test object was applied to a normal part of the body. Thus No. 1 found that when he thoughtlessly grasped a cup of very hot tea with his hand, his arm went into a violent extensor spasm and the cup was flung to the ground.

The mere fact, however, that a stimulus is potentially harmful is not in itself sufficient to ensure a response. In addition to a nocuous quality a stimulus, to be effective, must possess a certain intensity and duration. We found that for nociceptive reflexes of the upper limb in hemiplegia and in quadriplegia the threshold value of stimulus was usually lowest in the palm and axilla respectively. But here a single prick stimulus usually failed to evoke a response and often a series of pricks on one spot were unsuccessful. The most effective method of stimulation was by dragging the point of a pin along the skin, or by pinching the tissues. With moderately firm pressure a response usually appeared after the pin point had travelled about one inch and the distance required was less if the pressure on the pin was sufficiently heavy to break the surface. Hence at any one point within the receptive field summation of repeated pin-pricks was more capable of exciting a response than a single deep prick. But still more effective was serial stimulation of a number of points, as in dragging the point of the pin across the skin. The effectiveness of a stimulus depended therefore as much on the extent of the surface to which it was applied as on duration and intensity. This probably explains the effectiveness of a crushing stimulus such as a pinch or squeeze of the skin and deep tissues or firm pressure with a broad object such as the end of a pencil.

The direction of the movement evoked in the lower limb can often be altered in quadriplegia when the intensity of the stimulus applied to the thigh is changed. Thus, extension of the lower limb is evoked by drawing the tips of the fingers firmly along the skin on the inner aspect of the thigh, but the movement changes to flexion when the skin is firmly pinched. We have failed, however, to obtain reversal of direction of the reflex movement evoked by increasing the intensity of the stimulus, when it is applied to either the peripheral portion of lower limb in quadriplegia or hemiplegia or to any part of the upper limb in the latter. This was the case also in the upper limb in two examples of quadriplegia, but in No. 2 strong stimulation of the palm of the hand or inner part of the forearm by pinching usually excited a biphasic reaction in the upper limb, of which flexion at the elbow and

abduction at the shoulder formed the primary movement, and extension, adduction and internal rotation the secondary movement: stimuli of moderate strength applied to the hand or forearm evoked as a rule the extension reaction only.

In this patient and in No. 3 we did not succeed in evoking a flexion reflex of the upper limb by stimulation above the elbow, but a response of this kind was usually obtained in No. 4. In No. 1 also it could be evoked by the employment of a special form of stimulus. Thus, when the strongly tonic internal rotators of the shoulder were suddenly "stretched" by the observer, a movement consisting of flexion at the elbow and wrist, flexion and abduction of the fingers and extension and abduction of the thumb was evoked. It was always quickly replaced by extension, adduction and internal rotation of the limb.

(*B*) *Reflex Movements and Receptive Fields.*

Within recent years considerable attention has been directed to the study of reflex reactions evoked by potentially harmful stimulation of the lower extremities in animals and man after lesions of the central nervous system. Work at the bedside and in the laboratory has been inspired by the researches of Sherrington, who first described and analysed in detail the flexion reflex of the lower limb in decerebrate and spinal mammals. This phenomenon follows the application of a nocuous stimulus to the foot, and consists of flexion at hip and knee and dorsiflexion of the foot with reciprocal relaxation of the antagonistic extensors [10].

Several observers have investigated the reactions of the lower limb evoked by stimulating the foot under similar conditions in hemiplegia and paraplegia, and have found that they present in general the same characters as Sherrington's flexion reflex [6] and [16]. With lesions of the pyramidal tract at any level, pricking and scratching the sole of the foot evoke contraction of the flexors of the limb including the dorsiflexors of the foot and toes. As in animal preparations reciprocal relaxation of the extensors of the limb forms part of the response.

As a rule in hemiplegia and quadriplegia, when the resting posture of the lower limbs is one of rigid extension at all joints due to excessive tone of the extensor muscles, reflex flexion can be evoked by appropriate stimulation of any part of the limb below the knee. Although the response is most readily obtained, and with stimuli of least inten-

sity, from the outer part of the sole of the foot, yet by scratching or squeezing the tissues of the leg the reaction can usually be excited. The Schaefer, Oppenheim and Gordon toe-phenomena do not essentially differ from the reaction described by Babinski [1]. Each of these observers merely recommends stimulation of different parts of the receptive field for the flexion reflex, of which "extension" of the toes forms an integral part, or the employment of different kinds of nocuous stimuli.

In the disorders we are considering, therefore, the receptive field or reflexogenous zone for the flexion reflex has its focus in the sole of the foot, but also includes the whole of the peripheral portion of the limb below the knee. Above this level it is usual to obtain the opposite movement, namely, extension of the lower limb, by harmful stimulation of moderate intensity, such as pinching, pricking or scratching the tissues of the thigh. The extension reflex response consists of contraction of the extensors of the hip and knee and the muscles of the calf and sole of the foot. Along with the contraction of the extensors, the tone of the flexors of the knee also appears to increase if the reaction is evoked when the limb is extended. The receptive field has its focus in the groin, genital and perineal regions, but includes the thigh and extends sometimes for a short distance on to the abdominal wall, especially in the middle line.

Thus the hemiplegic or quadriplegic patient with spasticity of the lower limb in extension frequently exhibits two forms of reflex reaction to harmful stimulation:—

(1) Reflex flexion at all joints, including upward movement of the toes, most readily evoked from the sole of the foot, but also obtained by strong stimulation of the foot and leg below the knee and sometimes of the thigh; and

(2) Reflex extension of the limb, including downward movement of the foot and toes, excited by the application of appropriate stimuli to the proximal segment of the limb, especially to the parts lying near the perineum and external genitalia.

So far we have been dealing with phenomena which have already been described by Walshe [16], Riddoch [6] and others. Turning now to nociceptive reflex reactions of the upper extremity in severe spastic hemiplegia and quadriplegia, we have been able to evoke two main kinds of movements which displace the limb in opposite directions; in one reaction the limb is extended at the elbow and in the other it is flexed. As in the lower extremity, a response readily followed appropriate stimu-

lation within a large receptive field which extended from the peripheral portion to the base of the limb and even on to the trunk. But whereas in the lower extremity excitation of the leg and foot evoked reflex flexion, and of the thigh and neighbouring portion of the trunk usually reflex extension, in the upper limb in any one patient it was more usual to obtain one form of movement only, whether the exciting stimulus was applied above or below the elbow. In some of the cases, chiefly hemiplegic, flexion at the elbow, and in others, chiefly quadriplegic, extension at this joint occurred, but the receptive fields for both reactions were approximately the same.

In one quadriplegic patient (No. 2), however, either flexion of the elbow with abduction of the shoulder or extension of the elbow with adduction of the shoulder could be evoked by stimulation of the receptive field below the elbow; but appropriate stimulation above the elbow evoked an extension reaction only.

We propose to describe the first form of response as an "extension reflex of the upper limb." It was the response which constantly formed the fully developed nociceptive reflex reaction in three of our four quadriplegic patients, and in one patient with hemiplegia. It is well illustrated in the serial kinematograph pictures on p. 412. It is a vigorous movement which thrusts the extended upper limb backwards, at the same time producing extreme inward rotation, so that the palm of the hand at the height of the response faces outwards as well as backwards. The movement, when analysed at the different joints, is found to consist of elevation, adduction, retraction and inward rotation at the shoulder, extension at the elbow, hyperpronation of the forearm, flexion at the wrist and extreme adduction and extension of the fingers and thumb.

The movement is so rapid and strong that it was found to be impossible to determine by inspection and palpation the state of the antagonists while it was taking place—although they were certainly contracted while the position produced by the reflex was being maintained.

The receptive field for the reflex includes the palm of the hand and the palmar aspect of the fingers and thumb, the flexor and inner aspect of the forearm and upper arm, the axilla and the upper part of the chest, in front and behind, from about the second to the sixth ribs. The back of the hand and fingers and the posterior and outer aspects of the limb were usually unresponsive, and a fully developed reaction could not be excited from these parts. The most excitable portions of

the field were the four inner walls of the axilla and the palm of the hand, and the former more than the latter. That the axilla and inner aspect of the upper arm form the focus or part of the receptive field, where the threshold value of stimulus is lowest, is supported by the following observations:—

(1) It seems to be the part of the limb which is the earliest to respond to stimulation after the lesion of the spinal cord. For evidence on this point we are dependent upon observations in one case only made in No. 1 by Lieut.-Colonel Gordon Holmes five days after the injury. He noted that with the application of several pin-pricks in series to any part of the upper arm from the internal epicondyle to the axilla, a strong reflex movement occurred, consisting of adduction and inward rotation of the arm at the shoulder. By the time the patient came under our care, thirty-five and a half months after the injury, the receptive field had spread to its full extent.

(2) In all the quadriplegic patients who showed this reaction it could usually be evoked more easily from the axillary region than from the palm throughout the period of observation, which in No. 1 extended over six years. The difference in the thresholds for palm and axilla, however, was not great in any of our patients at a stage when their condition had become stationary.

(3) The minimal motor response from any part of the receptive field consisted most frequently of the movement which was the earliest reaction to appear after the initial spinal shock had passed away. In two of the patients the first movement to be evoked by stimulation of the upper arm or the chest was adduction and inward rotation at the shoulder, while in No. 3 the minimal response was extension of the elbow and flexion of the wrist and extension and adduction of the fingers.

The receptive field was strictly homolateral in all our examples of hemiplegia and quadriplegia, with one exception in which it was bilateral for the reflex in one upper limb. This patient (No. 3) was paralysed in all four limbs following a shell wound of the neck, but within two hours of his injury his right upper and lower extremities began to recover. By the time he came under our observation three days later all voluntary movements of his limbs on the right side could be performed, although his left upper and lower extremities were helpless except for feeble voluntary contraction of deltoid and biceps. Within a few months his right upper and lower extremities had, as regards motion, almost completely recovered. Analgesia of the inner

aspect of the upper limb however persisted. Return of voluntary power in his left upper extremity was slow, and the limb showed rigidity of the same kind and distribution as in Nos. 1 and 2. Reflex extension was obtained in his left upper limb only, but the receptive field was bilateral and practically symmetrical, comprising the palms of the hands, the inner aspect of both limbs and the upper part of the chest in front and behind from about the second to the fifth ribs.

The second form of nociceptive reflex reaction of the upper limb to which we draw attention in this paper was obtained in hemiplegic patients, and in two examples of quadriplegia (Nos. 2 and 4). Paralysis was, as in all our cases with one exception (No. 8), associated with spasticity, but in this group the rigidity preponderated in the flexors over the extensors of the upper limb.

The reflex response, for which a convenient term is "flexion reflex of the upper limb," was readily obtained in No. 5. On scratching the palm of the paralysed hand there occurred flexion of fingers, wrist and elbow, slight abduction and external rotation of the upper arm and elevation of the shoulder. These were the main components of the general response which varied in details according to the situation of the stimulus within the receptive field. This was invariably homolateral and corresponded almost exactly in extent with the receptive field for the extension reflex, covering the palmar aspect of the hand and fingers, the flexor and inner aspects of the forearm and upper arm, and spreading over the axilla and upper part of the chest on the same side. The focus of the receptive field, however, was differently situated in the two groups; it was peripheral in the palm for the flexor reaction in hemiplegia, and proximal in the axilla and upper arm for the extension reflex in quadriplegia. In the one example of quadriplegia (No. 4), however, in which flexion at the elbow was the most common response obtained at this joint, it was most easily elicited, as in the other cases of quadriplegia, from the axillary region.

Although flexion at the elbow in this patient usually formed part of the general reaction of the limb the response at other joints was different from that obtained in hemiplegia. For example, the movement evoked by scratching the palm or axilla consisted of adduction, elevation and outward rotation at the shoulder, flexion at the elbow and extension of the fingers and hand. This patient showed remarkable variability of response with alteration in the locality of the exciting stimulus, and the spasticity and paralysis of the limb were relatively slight; but a patient with a similar form of quadriplegia, who has just come under our observation, shows a reaction of the same kind.

In another example of quadriplegia (No. 2) with extensor rigidity of the upper limbs, flexion of the elbow with abduction and elevation of the shoulder could sometimes be evoked by strong stimulation of the tissues on the ulnar aspect of the forearm and the palm of the hand. The response was a quick unsustained movement which was followed by the more usual extension response. It was more readily evoked as a bilateral reaction by simultaneous stimulation of both upper limbs below the elbows.

All the hemiplegic patients examined did not exhibit the fully developed response. In many the reaction was limited to slight flexion of the fingers and hand with perhaps some pronation of the forearm, and in these the receptive field was confined to the palm. Thus the minimal motor response to harmful stimulation in hemiplegia with spasticity is often flexion of the fingers, and the focus of the receptive field the flexor aspect of the palm and digits. On the other hand in No. 7 we were able to observe the development of this reflex while the right upper limb remained completely paralysed and flaccid. The first movements to appear were abduction and external rotation at the shoulder accompanied at a later stage of recovery by flexion and slight supination at the elbow.

(*C*) *Influence of the Locality of the Stimulus on the Movements evoked—Local Sign.*

From the fact that one and the same nociceptive reflex in either the upper or the lower limb can be excited by stimulation of widely separate points within a large receptive field, it might have been assumed that considerable variations in the reactions evoked would be obtained. This, however, was not found to be the case except when the paralysis was slight as in No. 4. Although minor modifications were evident, depending mainly on the relative amount and duration of contraction in those muscles which were in most intimate nervous connection with the region stimulated, the general form of the response of the limb as a whole remained true to type. For example, when reflex flexion of the lower extremity was evoked by stimulating the outer part of the sole, with dorsiflexion of the foot there tended to be eversion, which was replaced by inversion and adduction when the locus of the stimulus was on the inner border of the foot. Again, in the reaction obtained by pinching the tissues round the knee, flexion at the knee and hip was sometimes relatively

greater than the movement of dorsiflexion of the foot and toes. But whichever part of the limb was stimulated within the receptive field for the reflex, flexion at all joints was the basic element of the response.

In the upper limb also, in cases of severe hemiplegia and quadriplegia, the nociceptive reactions evoked from the axilla and the palm of the hand were almost identical in general form. Variations attributable to the locality of the stimulus when present appeared in the initial part of the response before the general reaction developed completely. For example, pronation of the forearm and flexion of the wrist and fingers were early movements to follow stimulation of the ulnar part of the hand in quadriplegia, while extension and adduction of the fingers and thumb were usually evoked more easily from the thenar eminence. So also extension of the elbow and adduction and inward rotation of the shoulder were prominent when the stimulus was applied near the elbow and shoulder respectively. But as soon as the full reflex effect developed these differences disappeared, the complete reaction being the same from whichever part of the receptive field it was evoked.

Examples of severe hemiplegia showed in the nociceptive reaction in the upper limb a similar relative independence of the locus of the stimulus.

The reflexes we have been considering in the upper and lower limbs in hemiplegia and quadriplegia, therefore, belong to types. In each case appropriate stimulation of a large reflexogenous zone, which possesses a focus or central area where the threshold value of stimulus is lowest and a fairly well defined boundary, evokes a response which in form and direction of displacement of the corresponding limb-segments is almost but not absolutely stereotyped. With minimal reactions definite variations in the motor effects excited can be traced to the locality of the stimulus; but as soon as the complete or maximum reaction is evoked modifications become submerged and disappear.

Reduction in the adaptability of the reflex response to the site of nocuous stimuli has already been described in cases of paraplegia in flexion following transection of the spinal cord in the thoracic region. In these cases the prominent feature of the reflex picture is a massive reaction consisting of a bilateral flexor spasm of the trunk and lower extremities with evacuation of the bladder and rectum and excessive sweating. The receptive field comprises the whole of the paralysed region of the body and the situation of the stimulus has little effect on the form or extent of the movement except in sub-total reactions ([6] p. 345 and [4] p. 217).

There is one case in our series, however, in which reactions in the upper limb were influenced considerably by the situation of the exciting stimulus. The case No. 4 has already been touched upon in other connections and may profitably be referred to again in so far as it throws light on the present problem, but we hope to deal with it more fully in a later communication.

The patient at the age of 17 years suffered from an illness which was diagnosed as influenza. When he was allowed to get up after three weeks in bed, he noticed that his lower limbs were weak. The disorder slowly increased and within a year after the onset his left upper limb became affected in the same way. Later his right upper limb showed signs of impaired motor function. At the present time, three years after the attack of influenza, he shows moderate weakness and spasticity of all four limbs. He suffers little pain, and there is slight disturbance only of postural sensibility in his toes and of vesical and rectal functions. The functions of his cranial nerves are normal and radiographic examination of his spine fails to reveal any abnormality.

Muscular tone is definitely increased in his limbs and trunk, more on the left than on the right side, but not to any marked extent. In his upper limbs the excess of tone is about equal in all muscle-groups at the shoulders, at the elbows and wrists the flexors are more tonic than the extensors, but the fingers are hypotonic. In his lower limbs the extensors perhaps offer more resistance to passive movements than do the flexors.

From a large receptive field, comprising the whole upper limb the shoulder and the upper part of the chest, nociceptive reflex movements can be evoked. These reactions are most easily obtained with weak stimuli from the axillary walls and the inner aspect of the upper arm; for in this region a gentle moving contact on the skin with the tip of the finger is capable of evoking a movement of the limb. The back of the hand and the outer aspect of the forearm and upper arm are much less responsive.

The movement most commonly obtained consists of adduction and external rotation at the shoulder, flexion at the elbow and extension of the hand and fingers, but the response varies in a remarkable manner with alteration in the locality of the stimulus. Thus when the reaction is excited by scratching the palm of the hand, the movement at the shoulder is abduction and retraction of the upper arm. Stimulation of the back of the forearm yields adduction at the shoulder and of the

inner aspect abduction at this joint. Again, when the stimulus is applied to the skin over the deltoid, the response is mainly strong elevation of the shoulder with adduction and external rotation of the upper arm.

Without enumerating, for the present, further examples of reactions evoked in this patient, we wish to make clear that variability of response with change in the situation of the stimulus is an outstanding feature in this case, in which disturbances of voluntary power and muscular tone are relatively slight.

A comparison of the reactions in this patient with those in more severe cases of quadriplegia and hemiplegia justifies the conclusion that invariability of the response and enlargement of the receptive field for any one protective reflex are indicative of the extent to which disintegration of neural function has followed a lesion of the central nervous system; from this point of view they have the same significance as spasticity and paralysis.

The larger the field of a protective reflex the less do the reactions vary with the locality of the stimulus and the more restricted is the reflex behaviour of the limb as a whole.

(*D*) *Motor Phenomena accompanying the Reflex Movements of the Upper and Lower Limbs.*

Three of the quadriplegic patients (Nos. 1, 2 and 3) showed extensive reflex reactions involving the trunk and limbs.

Irradiation of reflex effect beyond the limb directly stimulated was very striking in Nos. 1 and 2, and was obtained with either reflex flexion of the lower extremity or reflex extension of the upper limb when the response was powerful. For example, in No. 2, the following reaction was evoked by applying a strong nocuous stimulus within the receptive field on the right upper extremity. The stimulated limb was thrust violently backward with adduction and inward rotation of the upper arm, strong extension and hyperpronation of the forearm, so that the palm of the hand faced outwards, flexion of the hand and extension and adduction of the fingers. On the left side all the muscles of the upper limb above the wrist were brought into action, but some more strongly than others, so that some displacement of the different parts of the limb occurred; there was slight adduction of the upper arm with forward movement of the shoulder, flexion and supination of the forearm and slight extension and radial deflection of the hand.

Palpation showed that erector spinæ and the anterior abdominal muscles contracted on both sides, and there was some flexion and rotation of the trunk to the right. Both extensors and flexors of the lower extremities became strongly contracted, the former more than the latter, so that there was rigid extension at hips, knees, and downward movement of the feet and toes; the right lower limb was more affected than the left.

The whole response obtained by stimulation of the palm of either hand in No. 1 was practically identical with that just described. There was augmentation of tonus in both prime movers and antagonists of the trunk, lower extremities and the crossed upper limb, and similar slight displacements at the different joints were observed. The only noteworthy difference was that the rotation of the trunk to the stimulated side was more prominent, and was accompanied by slight flexion at the hips when the patient was standing erect.

From this fairly uniform response, however, variations of some note occurred in the case of No. 3. It has already been mentioned that this patient was wounded by a piece of shell in the cervical portion of the spinal cord on July 15, 1916, and was for some time paralysed in all four limbs; but by March, 1917, his right upper and lower extremities had almost completely recovered. At this time the nociceptive extensor reaction was obtained in his left upper limb only. The reflex could, however, be excited by appropriate stimulation, not only from the palm and the inner half of the upper limb and the upper part of the chest on the left side, but also from the corresponding parts on the opposite side. Thus on scratching the skin of the right forearm, while no apparent change occurred in the muscles of the right arm, a typical extension response was evoked in the crossed upper limb. In this patient, then, there was identity in the forms of response excited by stimulating corresponding parts within the receptive field on the same and opposite sides. The motor effects produced by the activity of the reflex spread to the left lower extremity, and the direction of displacement noted on March 27, 1917, is another point of divergence from the reactions in Nos. 1 and 2. Whereas in the latter the remote results in the lower extremities of nocuous stimulation of an upper limb was invariably bilateral extension, in No. 3 they were unilateral and consisted of a flexor spasm of the left limb.

Scratching or pinching the toes or foot so as to excite strong reflex flexion of the lower extremity usually yielded results which were equally extensive. For example, in No. 2 a vigorous flexor reaction in

the left lower extremity was accompanied by the following changes in the musculature of the other three limbs and the trunk: Extension at all joints of the right lower extremity including downward movement of the foot and toes, but tonic contraction of the flexors (hamstrings and pretibial muscles) also occurred; vigorous contraction of the erectores spinæ and recti abdominales, the former more than the latter, and more on the stimulated side; in the left upper limb, slight but definite increase of tone in the adductors and abductors of the shoulder and in biceps and triceps with extension of the elbow; in the right upper limb, contraction of the adductors and abductors of the shoulder, biceps and triceps with adduction of the upper arm and slight flexion of the forearm.

A reaction of the same kind followed excitation of reflex flexion of the right lower limb. In this case triceps contracted more strongly than biceps in the homolateral upper limb.

Stimulation of the flexion reflex in the left lower extremity in No. 3 evoked as in No. 1 extension of the crossed lower limb, flexion at the elbow and adduction at the shoulder in the left (homolateral) upper limb and extension at the elbow, wrist and metacarpo-phalangeal joints in the right (crossed) upper limb.

In No. 1 the response in the crossed lower extremity and in the trunk was the same as in No. 2, but in the arms a stronger and more widespread reaction was obtained. Sometimes movements of all segments of the upper limbs were observed, and usually consisted of adduction of the upper arms, flexion and supination of the forearms, extension of the hands and of the fingers at the metacarpo-phalangeal joints with flexion at the interphalangeal joints. These displacements, however, were never extensive, especially at the proximal joints; and invariably palpation and inspection showed that, as in the associated reactions in No. 2, the antagonists of the muscles which brought about the displacements at the different joints did not become relaxed, as happened in the flexion reflex, but, on the contrary, were more contracted than before.

Summarizing our findings in three examples of quadriplegia, regarding change of attitude in the limbs and trunk effected by nocuous stimulation of a hand or foot, the following general conclusions are arrived at:—

(1) Stimulation of the hand or forearm evoked in Nos. 1 and 2 an extension response in the limb and adduction of the upper arm and flexion of the forearm in the crossed upper limb. To this rule

No 3 is an exception. In this case stimulation of the left hand or forearm gave the extension response which was not accompanied by evident changes in the crossed upper limb. Also stimulation of the right forearm evoked a reaction in the left upper limb only and at the elbow extension and not flexion was observed.

(2) The response in the lower limbs which followed appropriate stimulation of either upper limb in Nos. 1 and 2 was always bilateral extension at knee and ankle. In No. 3 only one experiment was made to investigate this point and in it a flexor spasm of the left lower limb accompanied an extension response in the left upper limb.

(3) Nocuous stimulation of the foot invariably evoked reflex flexion of the stimulated limb and extension of the crossed lower limb. In No. 2 the crossed response consisted of extension at hip and knee and downward movement of the foot and toes, while in No. 3 dorsiflexion of the foot and toes was sometimes observed.

(4) The remote effects in the upper limbs from excitation of reflex flexion in one lower limb were variable. In Nos. 2 and 3 the usual result obtained at the elbow was extension in the crossed limb and flexion in the homolateral limb. In No. 1 bilateral slight flexion was the rule.

Similar reactions were observed in our hemiplegic patients, but the response on the healthy side was comparatively feeble, especially in the upper limbs.

Sherrington has described the postural alterations in the limbs and trunk, which, in the decerebrate and high spinal animal preparations, follow nocuous stimulation of the fore and hind paws ([8] p. 165). They show a certain variability as regards the direction of displacement of the limbs in different preparations and in different experiments with the same preparation, but on the whole there tends to be a diagonal correspondence between fore and hind limbs. That is to say flexion of one fore limb is accompanied by flexion of the opposite hind limb and extension of the other two.

The order in which the different parts were involved in the reaction seemed to be the same in our patients as that described by Sherrington for animals. For example, stimulation of the right hand evoked movement of (1) the stimulated limb, (2) the trunk, (3) the right lower limb, (4) the left lower limb, and (5) the left upper limb. Sometimes, however, the crossed upper limb appeared to be innervated earlier than the lower limbs, but usually it seemed to be brought into action later and generally it was affected less than the rest of the body. As

regards the lower limbs the homolateral showed a stronger reaction than the contralateral.

With excitation of the flexion reflex of the lower extremity the sequence of involvement in the resulting reaction was: (1) the stimulated limb; (2) the opposite lower limb; (3) the trunk; (4) the homolateral upper limb; and (5) the crossed upper limb.

It was noticeable that stimulation of one lower extremity led to early and pronounced effects in the other, whereas in the case of the upper limbs the crossed reaction as a rule was relatively slight. Thus it would seem that the lower limbs are, as regards reflex connections, coupled together more closely than are the upper limbs. The former tend to react to reflex stimulation as a unit, while the latter are more independent of one another.

The sequence of irradiation, or order of spread, of reflex effect within each limb taking part in the associated reaction followed a definite rule which never varied. In the development of a complete reaction in association with a vigorous nociceptive reflex response of an upper or lower limb the "march" of innervation in each of the other three limbs was from base to periphery. Thus the muscles of the shoulder or hip contracted before a definite change could be noticed in those of the elbow or knee and these again before the muscles of the wrist or ankle. The field of musculature most affected outside the limb directly stimulated comprised the trunk and the proximal segments of the limbs. Increase in the intensity of the reaction led to: (1) augmentation of muscular contraction in these parts; and (2) progressive involvement of muscles more peripherally placed in the extremities.

It has already been pointed out that any displacement of the trunk and three limbs that occurred in the associated reaction was rarely extensive, although the muscles were frequently in strong contraction. This was undoubtedly due to the innervation of antagonistic muscles, which also explains another distinctive feature of the associated movements, namely, their slowness. The movements of the limb and trunk, in comparison with the rapid displacement of the stimulated limb, were carried out slowly, almost "deliberately." Each was a steady smooth performance, a gradual alteration of attitude rather than a movement. It was also generally uniphasic in the sense that for each segment the direction of displacement did not change in the reaction even when alternating movements of the stimulated limb were evoked by a prolonged stimulus. For example, by applying a continuous strong pinching stimulus to a toe, flexion of the lower limb was not usually main-

tained at a steady level, but was repeatedly broken through momentarily by active extension of the limb. The biphasic response thus evoked, however, was in general limited to the lower extremity under stimulation, the rest of the body maintaining a uniform posture.

(E) Reflex Movements in Relation to Tonic Postures of the Limb at rest.

Although almost all the muscles of the affected limbs were spastic, yet some were more hypertonic than their antagonists and so determined the postures of the limbs at rest. Thus in hemiplegia the upper limb is held in a position of flexion and the lower limb is extended. Extension was also the resting attitude of the lower limbs in the four examples of quadriplegia recorded in this paper, but while in one the upper limbs were slightly flexed, in the other three they were as a rule strongly extended at the elbows.

Extensor rigidity of all four limbs is the attitude usually observed in decerebrate mammalian preparations [12] and it may be helpful, for purposes of comparison with our own clinical observations, to summarize Sherrington's records of nociceptive movements in anthropoids in relation to the postural rigidity of the limbs ([9] p. 412).

Sherrington found that there is mutual antagonism between the results of nociceptive stimulation and the reflex posture of the limbs at rest, not only in the sense that the former are concerned with movements and the latter with maintained attitudes, but in the fact that muscles which are opposed in function are employed. Thus when the pad of the hind foot is pricked a movement of flexion of the limb is evoked and with contraction of the flexors of the knee and ankle there is simultaneous relaxation of the previously tonic extensors. A similar result follows harmful stimulation of the fore paw; when it is dipped into hot water flexion is evoked at the elbow, which at rest is tonically extended. He obtained results of the same kind in the decerebrate dog and cat. Nociceptive reflexes excited by stimulation of the peripheral portions of the limbs thus produce flexion at knee or elbow with reciprocal inhibition of tone in the corresponding extensors.

Innervation in the same limb by nociceptive reflexes of muscles which are opposed in function to those employed in postural reflexes, is however not a universal rule. There is at least one exception in the lower limb. In the decerebrate cat, dog and monkey, reflex flexion is the result which usually follows harmful stimulation of the greater part of the hind limb, but when the stimulus is applied to the thigh near the groin or to the perineum, extension and not flexion is evoked. Thus

the nociceptive reflex, which is excited from the base of the hind limb, employs the same muscles as are in tonic contraction when the limb is at rest.

So far as we know a nociceptive extension reflex has as yet not been described for the upper limb in laboratory preparations.

Let us now examine our clinical observations in the light of Sherrington's results in decerebrate animals. The postural attitude of the lower limbs in both hemiplegia and quadriplegia was extensor, due to preponderating tonicity of the extensor over the flexor muscles. This rigidity was inhibited and extension replaced by flexion when appropriate harmful stimuli were applied to the foot or to the leg below the knee. Sometimes reflex flexion could be obtained from the thigh by using intense stimuli. Usually, however, with stimuli of moderate strength extension and not flexion of the limb was evoked from the thigh, especially near the groin. Thus, in respect of the lower limb, the postural and nociceptive reactions correspond to those described by Sherrington in decerebrate mammals. Opposite effects result from harmful stimulation of the peripheral and basal portions of the limb. From the foot reflex flexion is evoked with inhibition of the tonic extensor rigidity and from the groin reflex extension. In the former case the nociceptive reflex employs the antagonists of the muscles which maintain the resting attitude of the limb, while in the latter the same muscle groups are innervated by the nociceptive and postural reflexes.

On making a similar analysis of the postural and nociceptive reactions of the upper limb, it is evident in the first place that, unlike the case of the lower limb, a single uniform resting attitude does not hold. Examining, for example, the posture at the elbow-joint in three examples of quadriplegia, there was as a rule bilateral tonic rigidity of triceps with maintained extension of this joint when the limbs were at rest. Occasionally, in one example (No. 2), after the limb had been kept flexed for some time either passively by the observer, or through the voluntary efforts of the patient, tonus was found to predominate in biceps. But this was not the usual state. In No. 4 spasticity was less developed and more equal in antagonistic muscle-groups, but it was always greater in the flexor at the elbow than in the extensor. A resting posture of flexion at the elbow is well recognized in spastic hemiplegia and is almost the invariable rule. The occurrence of one posture rather than another in these states is of course due to an unequal development of tonus in antagonistic muscle-pairs; in severe quadri-

plegia with well developed spasticity, the tonicity of triceps is greatly in excess over that of biceps, and in hemiplegia with rigidity the opposite state obtains. As regards the posture at the other joints of the limb in the two groups of cases there is less difference. In general the upper arm is adducted and rotated inwards, the forearm pronated and the wrist and fingers flexed.

In quadriplegia, with the exceptions of No. 4 and, under certain circumstances, No. 2, whose cases will be considered separately, the nociceptive reflex of the upper limb when excited generally brought into action those muscles which by their extensive tonic contraction determined the resting posture of the limb. For example, in Nos. 1, 2 and 3, the upper limb when at rest was held in the following position: Adduction and inward rotation at the shoulder, extension at the elbow, pronation of the forearm, and flexion at the wrist. Potentially harmful stimulation of the palm evoked a powerful movement of the limb, in which the upper arm was adducted and rotated inwards, the forearm extended and hyperpronated and the hand flexed. It must be again pointed out, however, that in No. 2 strong stimulation of the forearm or hand sometimes evoked a movement of abduction at the shoulder and flexion of the elbow; but it was quickly replaced by adduction and extension.

In hemiplegia, it is not the rule to find all the same muscles employed in nociceptive reflexes as in the postural reaction at rest. For example, the usual postural reaction at the shoulder is one of internal rotation and adduction, whereas the movement produced by nocuous stimulation is the result of contraction by the external rotators and abductors. Although flexion of the elbow, wrist and fingers is the general rule in both reflex reactions the movement of the forearm in the nociceptive reflex is always slight and in some cases tends to supination rather than pronation, the usual postural attitude of this segment of the limb at rest.

We have been comparing nociceptive reactions with postural reactions in cases of severe hemiplegia and quadriplegia in which spasticity and paralysis were well marked. One example of quadriplegia (No. 4) with much less spasticity and paralysis presented features of importance in relation to those which have just been discussed. In this patient the disturbance of function due probably to a high cervical lesion was altogether less than in the other cases of quadriplegia. All voluntary movements were fairly strong and free; sensation, apart from slight loss of postural sensibility in the feet and toes, was unaffected, and the vesical defect amounted to slight frequency of

micturition only; there were increased tendon-reflexes in the four limbs and the abnormal form of plantar response was present on both sides, but spasticity was relatively slight and was more evident in the lower than the upper limbs. Tonus however was quite definitely increased in all the affected muscles, but although it was, as in our other cases, unequally developed in antagonistic muscle-groups the inequality was not great. In the trunk and lower limbs, the extensors were somewhat more spastic than the flexors and there was difficulty in clearing the ground with the toes in walking. In the upper limbs the flexors of the wrists and pronators of the forearms resisted passive movements more than their antagonists and in this respect there was no difference from the distribution of tonus in the other examples of quadriplegia. But at the elbows the flexors and not the extensors were the more spastic muscles, and at the shoulders tone did not predominate in any one muscle group over another.

Nociceptive reflex reactions were easily excited in either upper limb especially from the axilla and inner aspect of the upper arm where a gentle moving contact with the tip of the finger was an effective stimulus. The reactions showed great variability with alteration of the site of the stimulus, but from the axilla the usual movement consisted of quick elevation, adduction and external rotation at the shoulder, slight flexion at the elbow and extension of the wrist and fingers. Flexion, however, was not the invariable movement at the elbow; extension usually occurred when the stimulus was applied to the posterior aspect of the forearm or upper arm near the elbow.

In this patient therefore in whom the paretic muscles were not excessively hypertonic the nociceptive reactions, which were determined by the site of the stimulus, showed no definite relationship to the distribution o muscular hypertonus or posture of the limb at rest.

Thus the correspondence between the muscles employed in the nociceptive reflex of the upper limb and those determining the resting posture is almost complete in many cases of severe quadriplegia, less intimate in cases of severe hemiplegia and practically absent in the case of slight quadriplegia we have just described.

On the other hand in all cases there were two forms of nociceptive reflex in the lower limb—one employing the muscles determining the extended posture of the limb at rest, the other bringing into action their antagonists.

These facts appear to show that nociceptive reactions are inde-

pendent of postural rigidity in the upper as compared to the lower limb, the independence being less marked the more severe the disintegration of function of the central nervous system.

(F) Motor Phenomena following Primary Reflex Movements.

Displacement of the limb in a particular direction is not the only consequence of the application of a harmful stimulus. The primary movement, after it has reached its height, is succeeded either by an active movement, which is opposite in direction to the first (rebound phenomenon of Sherrington ([8] p. 208), or by active maintenance of the attitude into which the limb has been placed by the movement, until gradual relaxation supervenes.

In cases of quadriplegia and hemiplegia with extensor rigidity of the lower limb, when the stimulus is applied below the knee, the former mode of reaction is usually observed, flexion followed by extension. This secondary movement of extension, however, is not a passive phenomenon in the sense of being due only to relaxation of the flexors and the weight of the limb. It is the consequence of active contraction of the extensors of the hip, knee and ankle and of the short muscles of the sole of the foot. Generally speaking, the strength of the secondary extensor contraction varies directly with (1) the strength of the primary flexion of the limb and (2) the degree of extensor rigidity present in the limb at rest.

Sherrington's analysis of similar biphasic movements of the hind limb in the decerebrate cat shows that during the execution of the primary movement of flexion the extensors are reciprocally inhibited. When the movement of flexion has reached its height and the flexors begin to relax, the extensors are innervated and the resulting contraction, which extends the limb, is stronger than the tonic contraction of these muscles before the reflex was excited (post-inhibitory exaltation). If with the limb at rest the degree of tonic contraction in the flexors is approximately equal to that in the extensors, as in the spinal cat or dog, then the secondary extension may be followed by flexion and this again by extension (reflex steppage movements).

This phenomenon of serial alternating reflex movements of flexion and extension can sometimes be obtained in cases of paraplegia ([7] p. 49) and quadriplegia in man when the degree of extensor tonus approximates that of flexor tonus in the lower limb (No. 4). But when hypertonus preponderates in the extensor muscles, as in hemiplegia and

most of our examples of spastic quadriplegia, then harmful stimulation of the foot evokes a single biphasic movement of flexion followed by extension, the series of alternating movements being interrupted at this point. The probable explanation of this event is that post-inhibitory exaltation of the flexor reflex arcs at the end of the extension phase of the movement is insufficient to overcome or inhibit the extensor hypertonus which becomes automatically reasserted.

In hemiplegia and quadriplegia, with extensor rigidity of the lower limb, reflex flexion evoked by the application of an appropriate stimulus is thus not maintained after the completion of the movement, but is inevitably followed by a movement of extension which brings the limb back to its usual position of rest. Although the reaction to a brief stimulus is limited to a simple biphasic movement of flexion followed by extension, a more prolonged stimulus is usually followed by a more complex response.

In this case a series of movements, extension alternating with flexion, separate the complete primary flexion of the limb from its final extension. These movements, however, are of small range and often of irregular rhythm. Although alternating movements of this kind may persist for a considerable period while steady stimulation is being maintained, they gradually die out and disappear, the limb being finally left in its previous state of tonic extension.

A reaction in which extension forms the first phase may also be evoked by stimulating the lower limb. This, it will be remembered, can be obtained by a stimulus applied to the thigh. If the stimulus is sufficiently strong, the primary movement of extension is followed by one of flexion, which again is succeeded by extension. The triphasic character of this reaction probably depends on the fact that, the initial movement being one of extension, three phases are necessary to bring the limb back to its resting attitude determined by its extensor rigidity. This may be compared to the biphasic result of stimulating the foot when the first movement is one of flexion and only a second is necessary to replace the limb in its extended position.

Active maintenance of the attitude into which a limb has been placed as the result of nocuous stimulation is the other kind of motor phenomenon to follow the primary movement. In the cases under consideration this occurs only when the primary reflex movement is one of extension of the limb and, therefore, is limited to reactions which commonly follow stimuli of moderate intensity applied above the knee. The prolonged contraction of the extensor muscles following a brief

stimulus must be regarded as an essential part of the reflex action. Relaxation takes place gradually after a few seconds and the limb is finally left with its original degree of extensor rigidity. In cases of spastic paraplegia with flexor rigidity this mode of reaction, represented by a prolonged contraction of the flexor muscles, is a common sequence of reflex flexion.

We have observed, therefore, two varieties of reactions following the application of a nocuous stimulus to the lower limb. The first is represented by alternating movements, which may be biphasic or triphasic according to whether the primary movement is one of flexion or extension. In the second there is a temporary maintenance of the posture produced by the primary movement.

As far as our experience goes nocuous stimulation of the upper limb usually fails to produce biphasic or triphasic phenomena following the primary reflex movement. According to the strength of the stimulus either a simple momentary displacement is as a rule evoked, or this new posture is maintained for a few seconds and then gradually fades away.

In hemiplegia, for instance, stimulation of the palm of the hand is followed by flexion, rarely by extension, of the elbow. In either case the after-effect of a stimulus of moderate intensity is to maintain for a time the displacement produced by the primary movement.

So also in quadriplegia, the same phenomena following the primary reflex movement are observed although the latter is more often extension than flexion at the elbow. In No. 2, however, a biphasic reaction in the upper limb could sometimes be evoked by strong stimulation of the flexor aspect of the forearm or hand. It consisted of flexion of the elbow and abduction of the shoulder quickly followed by strong extension and adduction.

The rarity in the upper limb of alternating movements such as occur in the lower limbs might at first sight be ascribed to the development of excessive tone in certain muscular groups. It is true that the muscles, which determine the direction of the reflex movements in either severe hemiplegia or quadriplegia, are generally those carrying a preponderating amount of tone when the limb is at rest. To this rule there are certain exceptions. For instance, in hemiplegia, whereas the adductors and internal rotators of the shoulder are more tonic than their antagonists at rest, abduction and external rotation are prominent features of the reflex movement. Moreover, it must be remembered that biphasic and triphasic movements can be evoked in the lower limb in spite of an

excessive extensor rigidity prevailing while the limb is at rest. Finally, we have been able to elicit a reflex movement without biphasic character in the upper limb of a hemiplegic patient, while his paralysed arm was completely flaccid and without tendon jerks.

A more probable explanation lies in the fact that the lower limb is primarily an organ for locomotion, a form of activity to which alternating movements are essential. On the other hand the inherent function of the upper limb is prehension, an action characterized by uniphasic flexion.

Chapter V.—Postural Reactions in Quadriplegia and Hemiplegia.

(*A*) *Nature of Spasticity.*

There can be no doubt that the phenomena which have been described in the preceding chapters are reflex in origin, although clearly they are not all of the same order. Judged by the effects produced it is possible to distinguish two groups, in one of which the reflex activities are expressed as postures, and in the other of which reflex movements involving displacement are manifested.

On the basis of this distinction it is our purpose now to compare some of the reflex reactions in quadriplegia and hemiplegia with those of a similar nature which have been investigated by physiologists in mammalian preparations and by clinicians in spastic paraplegia in man.

The state of maintained muscular contraction or rigidity which, under favourable conditions, follows the removal of the cerebral hemispheres in the monkey, cat, dog, and other animals, was first described by Sherrington in 1896 ([9] p. 411). Since then he has carried out extensive studies of the phenomenon, and has shown conclusively that it is reflex in origin and is postural in function—the posture which it subserves being the maintenance of the upright position of the body. In other words, it is reflex standing. In decerebrate rigidity he has described certain characteristic features the recognition of which has played an important part in the investigation of spastic states in man. Investigations along these lines have already been made upon the extensor rigidity of the lower limbs, which may follow lesions of the spinal cord ([16] p. 269 and [6] p. 329). In patients suffering from paraplegia of this form the lower extremities are rigid in extension from excessive developments of reflex tonus in the extensors of the hip, knee, and ankle (calf muscles). This rigidity has been found to

respond to tests in the same way as the rigidity of decerebrate mammalian preparations, and it appears to have the same functional significance. For when it is present the weight of the body can be supported in the upright position even in cases where voluntary power over the lower limbs is lost.

Hitherto, so far as we know, the spasticity of the somatic musculature in cases of quadriplegia has not been investigated in the light of Sherrington's researches, and the resemblance in most respects between the rigidity in our cases and that of the decerebrate animal is sufficiently striking to make the comparison a matter of some interest.

The distribution of the rigidity in the decerebrate preparation in the resting state is fairly constant, and determines the attitude of the body and its component parts. Sherrington ([9] p. 412) gives the following description of the posture of the decerebrate monkey when it is suspended in a cradle with its limbs hanging down. The fore-limbs are retracted at the shoulder-joints, straightened at the elbows and somewhat flexed at the wrists; the limbs are so rotated that the palmar surfaces of the hands face inwards. The hind-limbs are similarly retracted, extended at the hips, very stiffly at the knees, and to a less extent at the ankles. There is slight opisthotonos in the lumbo-sacral region, and the head of the animal is kept elevated with the chin tilted forwards.

The points of similarity between this attitude and the posture assumed by No. 1 in standing are in many respects very close. It is true that the slightly flexed attitude of the trunk in Nos. 1 and 2 is in contrast with the lumbar opisthotonos of the monkey, but this difference may be accounted for by the fact that the respective descriptions correspond to the erect position of the former and the horizontal prone attitude of the latter. It is in relation to the limbs, however, that the postures are most in agreement. The lower extremities are extended at all joints and the upper limbs adducted, internally rotated, but not markedly retracted at the shoulders, extended at the elbows, the forearms semi-pronated, and the wrists slightly flexed. The posture of the upper extremities is particularly arresting because of its rarity in man. Its occasional presence in the so-called tonic fits of Hughlings Jackson, however, is a fact to which Kinnier Wilson ([18] p. 220) has recently drawn attention. In the same paper this observer has described a state of maintained muscular contraction, following "acute physiological decerebration," in cases of internal hydrocephalus and of gross lesions of the mid-brain, which, as regards its distribution and the resulting

attitudes of the body and its component parts, resembles decerebrate rigidity. The postures of the limbs in some of his cases are reproduced with precision in our quadriplegic patients.

These considerations show that the distribution of the steady muscular tension or rigidity, to which in the absence of movement the maintained posture of the trunk and limbs in Nos. 1 and 2 was due, is at least very similar to those observed in animals after anatomical, and in man after physiological, decerebration. Broadly speaking, the attitude is one of general extension with pronation of the upper extremities or fore-limbs.

Sherrington states that the tone of those muscles which oppose the rigid muscles in decerebrate rigidity is mildly inhibited under the influence of reciprocal innervation. He says: "The tonic excitation is supplied only to one member, the extensor, of antagonistic muscle pairs. There is evidence that the flexors are under a mild tonic inhibition, the counterpart of the tonic excitation of the extensor motor neurons" ([10] p. 105). In other words the tonic or postural reflex, while bringing about steady contraction of the extensors, at the same time causes slight relaxation of the flexors, and the one process is proportionate in degree to the other although of opposite sign.

We made a point of repeatedly examining the condition of those antagonists which from their superficial position are open to palpation, and although our methods did not admit of exact measurements being made some trustworthy information was obtained. In the first place it was certain that the antagonists were less strongly contracted than the agonists. They were more pliable, yielded more to pressure and could be displaced from side to side more readily. In addition they offered less resistance to passive lengthening. We were able, moreover, to satisfy ourselves of the presence of a definite relationship between the states of tonus of antagonistic muscles. The strength of the postural contraction of the rigid muscles varied from time to time, and when the limbs were at rest the stronger the contraction in triceps the more relaxed the biceps and vice versâ. This point was well brought out by a comparison of the states of different antagonists. Some were found habitually to be more relaxed than others and it was noticed that those muscles which showed the least rigidity were opposed in their action to muscles which were invariably strongly contracted. Thus the curve that is given to the tip of the shoulder by deltoid in the healthy individual was absent in No. 1. The muscle was not appreciably wasted, but it was flat and more or less flabby in contrast to the

powerfully contracted and prominent pectorals. On the other hand when the upper limb was at rest triceps was rarely so rigid as the muscles of the anterior axillary wall and biceps usually stood out well, preserved its natural form and was of firmer consistence than deltoid.

Yet it could not be said that any of the antagonists of the rigid muscles which were examined were ever hypotonic; indeed at least some of them always appeared to be more strongly contracted than the corresponding muscles in a healthy individual. Notable examples were biceps, the extensors of the wrist, the hamstrings and the dorsiflexors of the foot. In this class should also be placed the muscles of the anterior abdominal wall which, according to Sherrington ([11] p. 212), exhibit the same reactions as the extensor muscles in the decerebrate animal. That the abdominal muscles subserve postural functions is shown by Kelling's experiments ([5] S. 161) on the intra-abdominal pressure in the dog.

This general but unequal raising of tonicity in somatic muscles when at rest we have also found to hold in spastic cerebral hemiplegia. Here the rigidity is distributed in the trunk and lower limb to the extensor muscles as in the extensor form of spastic paralysis from lesions of the spinal cord, but in the upper limb the flexors and not the extensors are overloaded with tone; hence the well-known attitude of the arm in such cases. But again, examination of the flexors of the lower extremity and trunk and of the extensors of the upper limb shows that these muscles maintain a steady contraction of a strength usually above the normal. As we shall see later, however, the rigid muscles, which by their overbearing activity determine the posture of the affected parts, show reactions which are poorly developed in their antagonists.

For a comparison of the spasticity in the cases under discussion with the rigidity of the decerebrate animal the relationship between the tonic states of resting antagonistic muscle-groups is a question of considerable importance. From laboratory experiments on animals Sherrington ([11] p. 196) has concluded that when one muscle of an antagonistic muscle-pair is contracted the antagonistic muscle is reciprocally relaxed. Our observations on the tonicity of the resting muscles are mainly in agreement with this statement. The muscles which are antagonistic to those exhibiting a high degree of tonic activity were relatively relaxed, but, compared with the muscles of the intact individual, their tonicity was increased.

With regard to the development of spasticity in different parts of

the body there is again considerable correspondence between our patients with quadriplegia and decerebrate animals. In the monkey, dog, cat, rabbit and guinea-pig rigidity after decerebration is more intense in the fore-limbs than in the hind-limbs ([12] p. 319). In the hind-limb the knee is the principal joint affected; the ankle is less implicated but is more rigid in extension than is the wrist in flexion. According to Sherrington the digits are never affected at all.

The condition found in Nos. 1 and 2, examples of quadriplegia, corresponds fairly closely with this description. In No. 1 the rigidity was much greater in the upper than in the lower extremities, although in No. 2 the distribution was less unequal. The extensors of the knees showed more tonic contraction than the extensors of the hips or ankles and, as in animals, the toes could be moved passively up and down without any resistance being encountered. Some of the muscles acting on the fingers, however, were hypertonic and others hypotonic, a condition which Sherrington does not describe in the decerebrate monkey. More especially in No. 1 the thumbs were steadily held against the palmar aspect of the index fingers, this position being maintained by powerful tonic contraction of the adductors and opposers of the thumb. Any effort on the part of the examiner to separate the thumb and index finger was met with strong resistance which increased within limits in proportion to the force exerted and to the rapidity of the attempt at passive displacement. Usually an adductor clonus was set up. But in contrast to the adductor rigidity of the thumb the fingers were extremely flexible. This was especially the case in No. 2 whose fingers could be passively hyper-extended and separated to an extreme degree. Passive flexion at the metacarpo-phalangeal joints could be less completely accomplished and there seemed to be quite definite increase of tone in the extensors of these joints. It will be remembered in passing that of the muscles which play directly on the wrist rigidity was more evident in the flexors than in the extensors.

The little and ring fingers in No. 1 also showed an absence of rigidity and an unusual freedom of movement. But while the other two fingers of the hand, in particular the index, could be readily over-extended, yet sudden jerky movements immediately excited brief contraction of the flexors. In the resting state and under the stimulus produced by slow stretching of their fibres the muscles of the radial fingers seemed to be almost flaccid, although a sudden increase of tension would excite a momentary resistance to passive lengthening.

A feature of postural activity in general, and one which is well

brought out in decerebrate rigidity, is the slow development of fatigue associated with it. If both hemispheres are removed and rigidity is well developed, strong tonic contraction of the extensor muscles is maintained for hours on end. This inexhaustible contractility was present in No. 1, especially in certain muscle-groups, notably the adductors and internal rotators of the shoulders, the pronators of the forearm and the flexors of the wrist. In No. 2, however, spasticity in the upper limbs was never so strong as in No. 1, and was maintained also at a less uniform level. When his arms were supported on the bed triceps usually became almost flabby, but tonic contraction could readily be excited again by passively moving the forearms backwards and forwards so as to alter the tension of the muscle. It should be remembered of course that in our cases conduction between the spinal cord and the cerebral hemispheres was by no means abolished; voluntary power and sensibility though disturbed were still present. This is of interest because in animals with hemi-decerebration, that is to say, with unilateral transection of the mid-brain, a similar waxing and waning of extensor rigidity is usually observed.

Perhaps the most characteristic phenomena of muscles showing postural contraction are what Sherrington has called the "lengthening" and "shortening" reactions. They can be demonstrated in the extensor muscles of the decerebrate preparation in the following way. With the animal lying on his side the observer elevates the hind-limb into the vertical position and the knee remains extended from tonic contraction of vasto-crureus. If he now passively bends the knee, so as to lengthen the extensor, the leg will be held in its new posture by the muscle against the action of gravity, the tonic contraction of the muscle being as strong as before. This is the "lengthening" reaction. So also when the attachments of vasto-crureus are approximated by passively extending the knee, the "shortening" reaction is evoked and the leg is retained in the position into which it has been placed by the same degree of steady contraction of the muscle.

Similar results were obtained from experiments of the same kind carried out on vasto-crureus and triceps in Nos. 1 and 2. In other words, they showed that (1) the steady tonic contraction of these muscles was sufficient to oppose the action of gravity on the part of the limb peripheral to the joint upon which each plays, and (2) this tonic or postural function is independent of the muscle length. This property of muscle in postural contraction to become reflexly adjusted to different lengths while counteracting the same load, led Sherrington to apply the term plastic tonus to what he later termed postural activity.

The conditions essential for the appearance of the shortening reaction are clearly shown in the case of the knee-jerk. When reflex postural activity is much diminished, as it is in the extensor of the knee, in spinal animals ([11] p. 211), and in spinal man ([6] p. 356), the knee-jerk may be brisk and ample, but relaxation is as quick or quicker than contraction, so that the leg falls like a dead weight. When, however, postural contraction is highly developed, as after decerebration, although the initial extensor contraction is just as rapid and easily evoked by tapping the patellar tendon as in the spinal preparation, relaxation is very different. In the decerebrate preparation the leg does not drop at once, but remains in the extended position into which the quick contraction of the knee-extensor has placed it, and is held there for several seconds; then it may gradually fall. A shortening reaction has been appended to the jerk contraction ([11] p. 211).

This phenomenon has been observed in the knee-jerk in spastic paraplegia in extension ([6] p. 357), and we found it to be remarkably developed in the patients described in this paper. In No. 2 it was particularly well shown in regard to both the triceps-jerk and the knee-jerk. The following observation was repeatedly made. The lower limb flexed at the knee and rotated outwards at the hip was passively raised off the bed and held in this position by the observer. Serial knee-jerks were then evoked by gently and rhythmically tapping the patellar tendon at intervals of several seconds. With each jerk the knee became a little more extended than before, and remained in this position until the next reaction was evoked. After several knee-jerks had been excited the position of the limb had become altered from one of flexion to one of almost complete extension. Knee-jerks evoked in this way thus lead to a step-like shortening of quadriceps extensor with progressive extension of the leg. The phenomenon was evoked equally well in No. 2 without the employment of percussion of the tendon. In his case sudden passive stretching of the extensor of the knee often excited a rapid clonus occurring at the rate of about 200 per minute. With each rhythmic movement the leg became extended a little more till finally extension at the knee was complete. The mechanism by which the extensor muscle was shortened in this case was the rapidly recurring excitation of the tonic reflex by the weight of the limb in each relaxation phase of the clonus. Progressive shortening reactions of the same order have recently been studied graphically in the vasto-crureus of the decerebrate cat by Viets ([15] p. 269).

The phenomenon, however, seemed to occur only when the jerks

evoked were of small amplitude. When the reflex was evoked by a strong stimulus so that the upward excursion of the leg was extensive the heel, in the relaxation phase, dropped almost immediately, sometimes to be arrested before it reached the bed. A reaction of this kind could be readily studied in No. 1, in whom extensor rigidity in the lower extremities was less well developed than in No. 2. We can best illustrate the main features of this reaction by describing briefly an experiment which was frequently repeated, and gave more or less constant results. With the patient lying on his back in bed his thigh was grasped by the observer, and his lower extremity suddenly elevated. The increased tension on quadriceps extensor caused by the weight of the leg and foot usually excited the tonic reflex, and the leg remained for a time extended in the air, although the extension was not, as a rule, complete. The patellar tendon or the leg was now smartly tapped so as to evoke a quick vigorous contraction in the extensor of the knee. This brought about sudden elevation of the leg, so that extension at knee became complete, followed almost immediately by rapid fall of the leg and foot from relaxation of the extensor muscle. If the thigh was being so held by the observer that there was considerable flexion at the hip relaxation of quadriceps extensor was usually arrested and replaced by tonic contraction before the heel reached the bed, and after one or two minor jerks the leg came to rest still suspended in the air, but at a lower level than before.

An analysis of the reaction shows that before the patellar tendon was tapped the leg was being supported against its own weight by the postural tonic contraction of quadriceps extensor. The appropriate stimulus for the knee-jerk, differing in its shorter duration and its more sudden application, excited a movement, namely, the upward jerk of the leg, and at the same time inhibited the tonic contraction of the extensor muscle. The quick contraction which caused the jerk was rapidly replaced by relaxation allowing the muscle to be passively stretched by the weight of the leg and foot; and it was only after the muscle had become elongated to more than its original length that the passive stretching excited once more the tonic or postural reflex. The descent of the limb was then arrested and the stoppage again evoked a mild jerk which was repeated before the reaction finally ceased.

The ease with which a tonic or postural reflex is inhibited by a kinetic or movement reflex employing antagonistic muscle groups has been shown by Sherrington and others to be one of the features of postural contraction. The phenomenon can be readily demonstrated in

the decerebrate preparation and in spastic paraplegia in extension in man by evoking a flexion reflex of the lower limb. With contraction of the limb-flexors there is simultaneous and proportionate relaxation of the previously rigid extensors. But in this respect the knee-jerk experiment we have just described is of particular interest, for it shows that in the same muscle inhibition of plastic tonus can occur simultaneously with the excitation of a quick contraction. Moreover the nature of the stimulation required for the postural and the kinetic or movement reactions is of the same kind although differing in intensity and duration. That dissociation of these reflex-effects can occur is most likely due to difference in reaction-times. Attention has been drawn to this difference in relation to the reflex reactions of the movement and postural adductors of the mollusc Pecten, and in this connection Sherrington ([11] p. 204) says that "the speed with which the postural muscle can by contraction shorten its length is relatively slow as compared with that of the movement muscle."

It is worthy of mention again that in our cases it seems that the strength of excitation of the kinetic reflex had some influence in allowing the dissociation to appear, for it was not observed when knee-jerks of small amplitude were evoked by gentle taps on the patellar tendon; then the shortening reaction was immediately appended on the twitch. The probable explanation of the difference obtained with variation in the strength of the stimulus is that, as in reciprocal innervation of antagonistic muscle-groups, the degree of inhibition of postural contraction is proportionate to the degree of excitation of kinetic contraction. Thus with a feeble knee-jerk inhibition of postural tonus in the vasto-crureus is slight, whereas it is more complete with a large knee-jerk.

While dealing with the distribution of rigidity in the resting muscles in the cases of quadriplegia we stated that although the antagonists of the rigid muscles were relatively inhibited yet, as compared with the resting muscles of the intact individual, they were hypertonic. It was also mentioned that certain properties which were conspicuous features of the agonists were poorly developed in the antagonists. These properties are the lengthening and shortening reactions.

When we say that the antagonists were hypertonic we mean that to the clinical tests we employed they behaved in the same way as the more tonic muscle-groups though in lesser degree. This was the case at all events with biceps and the hamstrings. For example, sudden passive stretching of these muscles led to a temporary increase in their tonic contraction, the increment disappearing when the stretching

process was stopped. Thus tonic reactions could to some extent be obtained in biceps and the hamstrings, but they were insufficiently developed to counteract the action of gravity on the limb and the greater tonic activity of the extensors. For in No. 1 when the upper limb was flexed at the elbow so that gravity acted on the forearm the tonic contraction of biceps was never sufficient to retain the forearm in this position and the elbow immediately became extended. The same result was obtained with the flexors of the knee.

As has already been mentioned, however, in No. 2 it was sometimes found that after his upper limb had lain in a flexed position across his chest for some time biceps became the more contracted of the antagonistic muscle-pair. When tested under these conditions the phenomena of well developed plastic tonus could be shown to be present; the forearm could now be placed in any position and held there by the tonic contraction of biceps. Progressive shortening of the muscle could also be induced by evoking a series of biceps jerks, although under the more usual influence of tricipital rigidity this reaction was not obtained.

The occurrence of plastic flexor tone in animal preparations is not unknown. According to Graham Brown ([3] p. 180), states of maintained flexion in the limbs may occasionally appear in the narcotized monkey during systematic examinations of cortical motor reactions. They may also appear from time to time in the decerebrate monkey or anthropoid ape, and maintained flexion of one fore-limb is often found in association with maintained extension of the other. (In our patient flexion of the upper limb occurred sometimes with flexion and sometimes with extension of the opposite upper limb.) Graham Brown notes that the posture of the limb may change either suddenly or gradually, a feature which we observed in No. 2. In No. 2 also, as in the monkey, triceps was in a state of mild relaxation so long as the plastic tonic contraction of biceps continued. In addition Graham Brown has found that in the monkey, which has been decerebrated by section of the neuraxis just in front of the anterior colliculi, unipolar stimulation on the cross section within an area corresponding with the site of the red nucleus evoked flexion of one fore-limb at the elbow with simultaneous extension of the other. These reactions are characterized by a marked after-discharge, so that for several minutes after cessation of the stimulus flexion is maintained on one side and extension on the other.

In hemiplegia with rigidity plastic tonus is usually distributed to

the flexors of the upper extremity, hence the well-known flexed, pronated attitude of the limb. Extensor rigidity is the rule in the lower extremity, but sometimes cases are observed, as in a patient recently under investigation in which the tonic contraction of the flexor predominated and the resting posture of the limb was one of flexion.

Although in No. 2 the upper limbs were sometimes found to be flexed at the elbow from alteration in the distribution of the plastic tonus, the rigidity in the lower extremities was invariably extensor. The flexors compared to the extensors were mildly relaxed; even when the limb was placed passively in a position of flexion the degree of tonic contraction in the flexors did not perceptibly increase. With the knee in various degrees of flexion experiments were carried out to determine whether the lengthening and shortening reactions could be obtained in the hamstrings. The tests employed showed that they were imperfectly developed and with some of the tests negative results were obtained. For example, when the hamstring tendons were tapped with the lower limb flexed and lying on its outer aspect on the bed a brisk contraction of the muscle was evoked, but the relaxation phase was rapid and even with strong stimulation the leg was not displaced. The elicitation of rhythmic hamstring jerks by repeated taps on the tendons at brief intervals yielded another interesting result: each twitch of the hamstrings appeared to be associated with simultaneous and temporary diminution in the tonic contraction of their opponent muscles, the extensors of the knees. This was the only additional phenomenon to be observed with the first three or four contractions. Corresponding in time to the relaxation phase of each subsequent hamstrings jerk, however, and following immediately on the stage of mild inhibition of the extensors, these muscles were felt to contract suddenly and a quick, short extension movement at the knee occurred. Repeated stimulation caused the leg to extend more and more in short steps.

This paradoxical phenomenon in which the knee is seen to be extended as the result of repeated taps on the hamstring tendon demonstrates the absence of an effective shortening reaction in the hamstring muscle. But this alone would not explain the steplike extension of the knee. It really depends on the operation of post-inhibitory exaltation, the knowledge of which we owe to Sherrington. The depression of one reflex which accompanies the excitation of its antagonist is followed by a phase of increased excitability. Applying this law to our experiment we find that after each of the first three or four taps on the hamstring tendon and the accompanying relaxation of

the vasto-crureus the only phenomenon to be observed is the re-establishment of tone in that muscle. During this time, however, the excitability of the extensor reflex arc had become exalted so that ultimately the re-establishment of extensor tone was in itself a sufficient stimulus to evoke an active shortening reaction of the muscle which was repeated after each subsequent hamstring jerk. Under conditions in which tonic contraction is approximately equally developed in the muscles innervated by antagonistic reflexes this process tends to provoke alternating reactions of opposite phase. For example, so long as stimulation is appropriate, reflex extension will be the inevitable consequence of reflex flexion and vice versa, as in the "mark-time" reflex of the spinal dog and spinal man. Successive induction ([8] p. 213) promotes biphasic activity. But where tonus is developed in one group of muscles in excess over the corresponding antagonists, as in the states of disordered function under consideration, rhythmic sequence of action in opposing muscles is damped down or prevented by the predominant rigidity which overcomes the effect of post-inhibitory exaltation on the reflex arcs controlling the less tonic muscle-groups.

On analysis the resemblance between the rigidity in quadriplegia in extension and the maintained tonic contraction in the decerebrate animal preparation suggests that the two states are closely related. Attention has already been drawn by Walshe ([17] p. 6) to the similarities between the spasticity of hemiplegia and of decerebrate rigidity, and our observations in general support his conclusions.

With Sherrington we believe that tonus is a postural reaction and that, in the states under consideration, it is reflex in origin, the stimulus being provided by tension acting on receptors within the muscles themselves and their tendons. The natural stimulus which evokes the reaction in the trunk and limbs, and so determines the resting attitude of the body, is provided by the tension exerted by gravity, and Sherrington has interpreted the decerebrate rigidity found in extensor or anti-gravity muscles in animals as reflex standing.

Is it reasonable to offer such an explanation for the rigidity or postural contraction in the upper limbs? Man is bipedal and the freedom of the upper limbs from the bondage of locomotion allows them to be used as organs for exploration and prehension. The grasping and conveyance of food to the mouth is probably the elementary function of the human hand and arm. A moment's reflection, however, will show that the upper extremities, although they have ceased to take any direct part in locomotory acts, are still bound to the lower extremities in the

automatic movements of walking and running. In these movements the arms may be brought into action in two different ways; they may perform rhythmic biphasic movements or they may assume postures. The former mode of activity is observed in walking and the latter in running. Thus, when a man is walking quickly along the street he involuntarily swings his arms, the timing, vigour and amplitude of the movements corresponding closely to those of the lower limbs. The arms simultaneously move in opposite directions and the direction of movement of the four limbs at any moment is such that there is a diagonal correspondence. Thus the right upper and left lower limbs are coupled together and swing forwards or backwards at the same time, flexion of one arm occurring with extension of the leg on the same side. The movements of the limbs in walking are therefore the same in man as in four-footed mammals, those of the upper limbs being indirectly locomotory in the former and directly in the latter.

In the act of running, however, postures rather than movements tend to be adopted in the upper limbs. The attitude most commonly observed in a runner going at full pace is one of adduction at the shoulders, flexion at the elbows, flexion of the fingers and extension of the wrists. All the muscles of the upper limbs are in contraction and although to-and-fro movements at the shoulders in diagonal correspondence with the movements of the lower limbs are observed they are usually small.

Let us consider these observations on locomotion in the normal individual in relation with the phenomena we have been investigating. The flexed attitude of the upper limb in relation with the extended lower limb in hemiplegia might be held to have a locomotory significance. This may in part be true, and certainly many hemiplegic patients when walking automatically swing their paralysed arms as they move forward their unaffected lower limbs. But taking into consideration the fact that the hemiplegic patient as a rule recovers the power of voluntary flexion better than that of extension of his upper limb, the rigidity of the flexors is probably more intimately connected with the prehensile function of the limb.

It is otherwise, however, with the upper limbs in quadriplegia in extension. They are kept extended at the elbows and if voluntary power is not abolished extension is stronger than flexion. Let us briefly review again the general attitude of the patient in standing. His lower limbs are straight with his feet apart; he stoops a little forward from slight flexion of his spine and hips; his upper limbs hang down a little

in front of his trunk, extended at the elbows, the forearms somewhat pronated, the wrists slightly flexed; his fingers are extended at the metacarpo-phalangeal joints and otherwise flexed. The whole attitude in its general features is not unlike the position often adopted by the higher apes in standing and suggests in grotesque fashion a static posture in a stage of human development when the upper limbs may have subserved more direct locomotory functions.

Disease or injury of the nervous system results in disturbance of function. There is destruction of activities which in the history of the individual are recently acquired, while those which are older, more organized and automatic are preserved in an altered form. These positive manifestations of disintegrated nervous activity are unusual and abnormal in so far as (1) they are excessive developments of normal phenomena released from the physiological control to which they have been subjected by the intact nervous system; and (2) they present features bearing less resemblance to normal reactions in proportion to the extent to which the functional activities of the part have been acquired. Thus the flexion reflex seen in paraplegia is little more than a crude exaggeration of a normal reaction, especially in infants, belonging to a limb which is largely automatic in its function. On the other hand, the reflex activities displayed by the upper limb in quadriplegic patients may diverge widely and strangely from those usually associated with a part which has been educated up to an infinite variety of specialized movements.

The reactions laid bare by injury or disease cannot be looked upon as exact reproductions of modes of behaviour evoked under similar conditions at an earlier stage of human development. In spite of their modifications, however, they still bear traces of their ancestry, and also of their relationship with analogous reactions in the intact individual.

(*B*) *Significance of "Associated Movements."*

Much has been written on the subject of associated movements, particularly by French clinicians; but, as Walshe ([17] p. 16) has pointed out, little attempt has been made to investigate them in the light of modern physiological knowledge. We have studied these reactions in a number of hemiplegic and quadriplegic patients, and believe that our observations justify certain conclusions which seem to throw some light into the obscurity surrounding this subject.

Since reflex reactions of widely different kinds have been erroneously

included under the heading "associated movements," we will define our use of the term in order that there may be as little misunderstanding as possible. "Associated movements" are automatic activities which fix or alter the posture of a part or parts when some other portion of the body is brought into action by either voluntary effort or artificial reflex stimulation. Closely related with such reactions are the muscular activities which occur in the limbs and trunk in association with such automatic acts as yawning, "stretching," and coughing.

The term was originally employed for movements of the paralysed limbs in spastic hemiplegia, which occur with strong voluntary movements of the unaffected limbs. But our observations on reactions of this kind show that the response is not, as a rule, confined to the paralysed half of the body, but also involves the non-paralysed parts. Thus when No. 5, an example of right hemiplegia, attempted voluntarily to bend his left elbow against resistance the following movements took place in his trunk and other three limbs. His right (paralysed) upper limb became adducted and rotated inwards at the shoulder, incompletely extended at the elbow and fully at the wrist and the hand clenched. His trunk became slightly extended from contraction of the muscles on both sides, and his lower limbs stretched out.

A general reaction could be obtained not only when he tried to move one of his non-paralysed limbs against resistance, but also when he made strong voluntary efforts with his paretic upper or lower limb on the right side. For example, with voluntary flexion of his right elbow, the general movement evoked consisted of slight extension of his trunk, flexion of his left elbow and bilateral extension of his lower limbs. Again, voluntary extension of his right (paralysed) lower limb evoked extension of his other three limbs.

But movement is not the main part of associated reactions of this kind, in spite of the prominent place given to it by the majority of previous observers. The most striking feature of an automatic and associated response evoked by strong voluntary effort to displace a part of the body, whether paretic or not, is increased contraction of antagonistic pairs of muscle-groups in the trunk and other three limbs, with the result that strong fixation rather than movement of parts is obtained. Sometimes, but by no means invariably, one group of muscles acting on a joint are contracted more strongly than the opponents, so that movement at this joint results. But such movement is slow, and its appearance seems to be determined by the strength of the voluntary effort made by the patient.

Undoubtedly the earliest and most constant response is increased contraction of all the muscles acting on a joint. This appears first in the muscles of the trunk and of the proximal joints of the limbs, and progressively spreads towards the fingers and toes with increase in the intensity of the exciting stimulus. The associated reaction is always strongest and involves most muscles in the paralysed limbs, and in them appears earlier than in the healthy upper and lower limbs. Thus when No. 5 tried to bend his left (unaffected) lower limb against resistance the associated reaction was observed first in his right lower limb and trunk, then in his right upper limb, and lastly in his left upper extremity. Also, in his trunk and paralysed limbs all the muscles were involved, and movements at different joints were observed at a stage in the reaction when in his left upper limb the muscles of the shoulder only showed increased contraction without actual displacement at this joint.

Associated reactions of the same order as those we have just described in hemiplegia were evoked in the trunk and three limbs in patients with quadriplegia when strong voluntary movements were attempted in the fourth limb against resistance. In No. 2 voluntary power was more impaired in the limbs of the right than those of the left side. Resisted voluntary flexion of the left elbow excited the following response: In the right upper limb there was elevation adduction and backward movement at the shoulder with slight flexion at the elbow and wrist; in the trunk the homolateral rectus abdominalis and contralateral erector spinæ were brought into action more than their antagonists, so that the trunk was rotated to the right; the lower extremities were rigidly extended.

The reaction obtained when the patient attempted to extend his left elbow against resistance was as follows: In the right upper limb adduction and downward movement occurred at the shoulder, extension of the elbow, some pronation of the forearm and slight extension of the wrist; the trunk rotated to the left and the lower limbs became extended at all joints.

Two further observations only will be quoted from our records of this patient's reactions. With strong voluntary flexion of his left knee his right first flexed and then strongly extended with downward movement of the foot; his trunk arched forwards with rotation to the left and his upper limbs rotated inwards and abducted slightly at the shoulders, and flexed at the elbows and wrists.

Attempted voluntary extension of his left knee against resistance

evoked a response in the trunk and opposite lower limb which was very similar to that just described, but in the upper limbs there was adduction with internal rotation at the shoulders, strong extension at the elbows, pronation of the forearms and slight flexion of the wrists.

But, as in hemiplegia, these movements, which under similar conditions varied to some extent even in the same patient in different experiments, did not form the earliest part of the response nor did they necessarily appear, especially in mild reactions. They really were an index of the intensity of the exciting stimulus, namely the voluntary effort of the patient to move one of his limbs. In a reaction the sequence of events was not difficult to follow. If, for example, the patient squeezed an object in his hand all the muscles playing on the wrist, elbow and shoulder were automatically brought into action, although with the exception of extension of the wrist no actual movement need occur at these joints. As he increased his efforts to squeeze the object in his hand the associated response spread from the limb directly stimulated and became manifest first in increased contraction in all the muscles of the trunk and proximal joints of his limbs. The other upper limb and the lower limb on the stimulated side were involved before the crossed lower limb. Still stronger stimulation resulted in spread of the muscular field involved towards the peripheral parts of the limbs and in movements at some of the joints due to greater contraction of some groups of muscles over their antagonists.

Reactions similar to these described above can also be excited by strong protective reflex movements in an upper or lower limb in both quadriplegia and hemiplegia. Thus in No. 2, when the extension reflex was excited in the left upper extremity, the right upper limb became more adducted at the shoulder and slightly flexed at the elbow; all the muscles of the trunk, but especially those of the left side, became contracted and strong extension occurred in the lower extremities, in the left more than the right.

Again with powerful reflex flexion of the left lower limb there occurred extension of the right lower limb at all joints, strong contraction of the erectores spinæ and to a less extent of the rectus abdominalis and oblique muscles on both sides and bilateral contraction of the abductors and adductors of the shoulder and flexors and extensors of the elbow.

In No. 5, an example of right hemiplegia, the following associated reactions occurred with strong reflex flexion of the right upper limb:

Contraction of all the muscles acting on the left shoulder and elbow joints, although no movement actually took place; all muscles of the trunk and of both lower limbs were brought into action, more powerfully on the right than the left side. Owing to the predominant contraction of the extensor muscles in the lower extremities these limbs remained in an extended position.

With reflex flexion of the right lower limb there occurred contraction of all the muscles of the left lower limb, which remained firmly extended; in the right upper limb contraction of all muscles on both sides of the trunk, strong contraction of abductors and adductors of the shoulder, of the flexors and extensors of the elbow, with a slow and incomplete movement of extension, of the flexors and extensors of the wrist with extension of this joint and of the flexors of the fingers so that the hand became clenched; in the left upper limb contraction of all the muscles of the shoulder and elbow-joints without any movement of the limb.

The remote reflex phenomena we have been describing, whether in association with powerful voluntary contractions against resistance or with strong protective reflex reactions in a limb, possess certain points in common. In the first place, the outstanding feature of the widespread response was increased tone in prime movers and antagonists in regions of the body remote from the stimulated limb. That is to say, although some of the limb segments and the trunk were often displaced the movement was due not to unopposed contraction of certain muscles but rather to an excess of contraction in some muscle-groups over that of their corresponding opponents. Whether a movement occurred or not accession of tone in all the muscles acting on a joint formed the earliest and most constant part of the associated reaction.

Secondly, it was necessary that the protective reflex response or the voluntary muscular effort should be vigorous before actual movements were obtained in the other limbs or trunk, and any displacement was usually of small extent and slow in its development.

Thirdly, these associated reactions invariably appeared earlier, spread farther and were more intense in paralysed parts, although as the result of a strong stimulus they involved healthy regions of the body as well.

Lastly, the muscles first innervated in a limb were those which played on the proximal joints, while any displacement was generally more noticeable in the peripheral segments.

To sum up, our observations seem to show that the phenomena

usually spoken of as associated movements are in reality widespread postural reactions. This conclusion has also been arrived at by Walshe ([17] p. 19), but the prominent feature of these reactions, namely tonic innervation of opposing muscle-groups, has hitherto escaped attention. When one functional muscle-group acting on a joint is innervated more strongly than its antagonists movement of the limb results but it is leisurely and rarely extensive. Movement, however, does not occur and a fixed posture is maintained when the antagonistic muscles are almost equally contracted and balance one another in their action. The muscles mainly involved are those of the trunk and the proximal segments of the limbs. These reactions resemble in their main characteristics those observed in the normal individual when strong voluntary efforts are made to move a limb against a heavy load. Under such circumstances the body is automatically adjusted for static purposes in such a way that the voluntary effort is directed towards one part of the body under the most favourable mechanical conditions, whilst at the same time equilibrium is maintained.

Chapter VI.—Distribution of Innervation and Inhibition in Reflex Movements and Postural Reactions.

Beevor's investigations ([2] p. 54) on muscular action have demonstrated that in regard to their behaviour in a voluntary movement of a limb muscles can be classified in two groups: (1) those which are contracted and (2) those which are relaxed. Of the muscles which show increased contraction some perform the function of prime movers and are directly concerned in bringing about the required movement at a joint while the others carry out subsidiary functions as fixators and synergists. Muscular relaxation is observed in those muscles only which by their action would prevent the desired movement from taking place; in other words they are the antagonists of the prime movers. Thus, when voluntary flexion of the elbow is carried out against resistance, all the muscles acting on the shoulder and wrist joints may be thrown into contraction as fixators or synergists while the inner and outer heads of triceps, the true antagonists of biceps and supinator longus, are relaxed.

When the movement of flexion is confined to the elbow-joint the state of the long head of triceps is of considerable interest. Its behaviour is opposed to that of the humeral parts of the muscle and it contracts as a fixator of the shoulder. Thus, triceps, which is

generally considered as an anatomical unit with one function, namely extension of the elbow, is composed of two parts which may subserve different functions in a flexor movement of the limb. Both parts have a common insertion into the olecranon process and may act together as extensors of the elbow. But the humeral portion is the only pure extensor, and as the true antagonist of supinator longus and the short head of biceps is relaxed in flexion of this joint. The long or scapular portion of triceps crosses the shoulder as well as the elbow-joints, and in voluntary flexion of the elbow may be reflexly brought into action, not as an extensor of this joint, but with long head of biceps to fix the shoulder and so facilitate the short head of biceps in flexing the elbow.

Several other limb muscles, such as biceps and rectus femoris, belong to the same group as triceps, in that each is physiologically speaking made up of two muscles with a common insertion but different origins and under certain circumstances subserving different functions. One part acts on one joint only, the other acting on two, and while both may be contracted or relaxed together under other conditions there may be simultaneous innervation of one and inhibition of the other. Ignorance of this fact has led certain observers to deny the occurrence of muscular relaxation in reflex movements ([14] p. 154).

In voluntary movements of the paretic limbs in quadriplegia and hemiplegia the innervation of muscles directly or indirectly concerned obeys the same laws as those obtaining in voluntary movements in healthy individuals. With increased contraction of prime movers, synergists and fixators there is relaxation of pure antagonists. For example, with voluntary abduction of the arm at the shoulder against resistance the pectorals, which in the resting posture are strongly contracted, undergo relaxation.

The motor taxis in reflex movements of the limbs in decerebrate and spinal mammalian preparations has been analysed by Sherrington [8], who has brought to light most of the known physiological laws of motor innervation. In a movement such as reflex flexion of the fore or hind limb certain muscles become contracted as prime movers, synergists, and fixators, while others, the true antagonists, such as vasto-crureus, are relaxed. The muscles of the first group show concurrent innervation, and those of the second group reciprocal inhibition. Further, the degree of contraction in the fixators and synergists, and of relaxation in the antagonists, is determined by

and proportionate to the amount of innervation of the prime movers.

Our investigations on the examples of spastic quadriplegia and hemiplegia quoted in this paper seem to show that reciprocal innervation of antagonistic muscles is present in some of the reflex movements examined. For example, when the flexion reflex of the lower limb is excited by appropriate stimulation, vastus internus, which with the limb at rest is in strong tonic contraction, becomes relaxed. The tightening of the patellar tendon that invariably occurs is due to the action of the long head of rectus femoris which is employed in the reflex as a flexor of the hip. Sometimes the muscles of the calf contract, although less strongly, than the dorsiflexors of the ankle; but it should be remembered that the only pure extensor of the ankle, namely soleus, is covered over by gastrocnemius, which, by its attachment to the femur, may act under certain circumstances as a flexor at this joint.

In the flexion reflex of the upper limb in hemiplegia the tone of at least some of the antagonists is diminished. The movement at the proximal joints consists of abduction with external rotation of the shoulder and flexion of the elbow. The pectorals thus act as opponents, and since they can readily be palpated, and are in the resting posture of the limb strongly contracted, it is easy to determine that relaxation occurs. After carrying out many investigations in different patients on the state of triceps during a flexor reaction of the upper limb, we arrived at the conclusion that it sometimes became relaxed and at other times its tone seemed to increase. So far as it could be ascertained, the behaviour of the muscle depended upon whether or not movement occurred at the elbow and shoulder. If biceps and supinator longus were brought strongly into action without consequent displacement of the elbow, and if the shoulder remained fixed, then triceps, in part at least, was also innervated. But with strong flexion of the elbow the humeral portion of triceps seemed to relax. It was difficult to be certain what occurred in the extensors of the wrist and fingers when flexion was evoked by scratching the palm of the hand.

Reciprocal relaxation of antagonists to the prime movers could also be demonstrated in the reflex response with flexion of the elbow in Nos. 2 and 4, examples of quadriplegia.

So far, then, it may be said that in those reflex movements which flex or fold up the upper or lower limb in quadriplegia or hemiplegia, those movements which can be evoked from the periphery of the

corresponding limb, there is concurrent innervation of synergists, fixators and prime movers and reciprocal inhibition of at least some antagonists. The motor-taxis evident in these reactions, therefore, corresponds to that in voluntary movements against resistance and in reflex flexion of the fore and hind limbs in decerebrate and spinal animals.

When we come to analyse the distribution of innervation which occurs during the extension reflex of the upper limb in quadriplegia, we are at once faced with the difficulty of ascertaining what takes place in all the muscles antagonistic to the prime movers.

It will be recollected that in the cases of quadriplegia (Nos. 1, 2, and 3) the upper limb presented general spasticity, which was more marked in those muscles determining the resting posture, consisting of adduction and internal rotation at the shoulder, extension at the elbow, pronation of the forearm, slight flexion of the wrist and of the fingers at the interphalangeal joints. This resting attitude was accentuated during the extension reflex with the addition of strong retraction at the shoulder and hyperextension of the fingers. While it was impossible to be certain whether muscles antagonistic to the prime movers were actively contracted or not during the development of the movement, it was comparatively easy to ascertain their condition during the maintenance of the posture produced by the reflex. For example, there was no doubt that deltoid, spinati, and latissimus dorsi were strongly contracted as well as pectorals, and that the tone of biceps increased as well as that of triceps. More difficulty was experienced in determining whether the extensors of the wrist were in a state of tonic contraction owing to the obvious activity of the extensors of the fingers.

There are similar obstacles to precise observation with ordinary clinical methods on the state of certain muscles of the lower limb when its extension reflex is excited, but it is justifiable to draw attention to the resemblance presented by the latter to the extension reflex of the upper limb. In both cases stimulation of the reflex results in accentuation of the already extended attitude of the limb. Although it is not justifiable to state that no muscles undergo relaxation it is certain that the majority of the muscles accessible to palpation, whether fixators or antagonists, are concurrently innervated. The effect of the reflex is in fact more in the nature of the assumption of a posture than of a movement comparable to the withdrawal of the limb brought about by the flexor reflexes.

It should also be noticed that the focus of the receptive field for the extension reflex in both the upper and lower extremities is found at the base of the limb.

Concurrent innervation of antagonistic muscles is perhaps most easily demonstrated in the remote postural reactions which are frequently associated with either voluntary movement against resistance or a strong reflex response in a limb. These reactions have been described in detail in the preceding chapter, and stress has been laid on certain characteristic features observed in them. One of the most noticeable and constant was the simultaneous increase of tone in all the muscles acting on any joint involved in the reaction, irrespective of the tonic state of the muscles at rest.

Sometimes the innervation was apparently almost balanced and the joint was more or less fixed; at other times, and more commonly in some joints than in others, and depending also upon the strength of the response in the limb directly stimulated, innervation was unequal and displacement occurred from stronger contraction of some muscles than of others. Usually the displacement was slow and did not alter in direction with increase of intensity of the exciting stimulus. But occasionally reversal of movement was noticed. Thus in No. 2, an example of quadriplegia, with increasing voluntary effort to bend the left knee against resistance, slight flexion of the right lower limb usually appeared early but was later replaced by extension at all joints.

An interesting example of a similar condition is provided by the coarse tremor of the hand that was frequently observed in associated postural reactions in the upper limb in quadriplegia and also in the paralysed upper limb in hemiplegia. The posture of the wrist with the upper limb at rest in both cases was slight flexion due to preponderance of tone in the flexor muscles. But in an associated reaction evoked by either voluntary or reflex movement of another limb the distribution of tonus was changed. Contraction in both muscle groups increased, but the accession of tone was greater in the extensors than in the flexors, so that the posture of the wrist gradually changed from one of flexion toward one of extension. A point in the reaction was reached when the slow extension of the wrist was replaced by a coarse tremor, flexion alternating with extension. In the case of the paralysed upper limb in hemiplegia if the associated postural reaction included extension at the elbow and supination of the forearm tremor of these parts was also liable to occur at a certain stage in the response. It could, however, be made to disappear by either diminishing or increasing the intensity of

the reaction, which had the effect of allowing the contraction of the flexors of the wrist and elbow and the pronators of the forearm to be greater or less than their corresponding antagonists. As will become evident, this is a point of some importance in the interpretation of the physiological significance of the tremor.

The probable explanation of this biphasic reaction is found by referring to Sherrington's investigations on rhythmic reflexes of the lower limbs in decerebrate preparations with extensor rigidity ([13] p. 96). His experiment was as follows: Faradic stimulation of either peroneal nerve evoked flexion of the limb and extension of the other lower limb, the antagonists in both cases being reciprocally relaxed. When, however, both nerves were simultaneously excited with faradic stimuli of approximately the same strength relaxation of antagonists disappeared and opposing muscle groups of both knees became contracted. Along with this change from relaxation to contraction in the antagonists the degree of contraction of either the flexor on the stimulated side or the extensor on the opposite side was found to be less than when one peroneal nerve only was stimulated. He was able to show that the innervation of each muscle group under bilateral stimulation was equivalent to the algebraical summation of the two opposing influences. Thus for example the flexor of the right knee was strongly contracted when stimulated through the right peroneal nerve only, but the degree of this contraction was diminished when the left nerve was simultaneously excited, the effect of which as regards the muscle was relaxation. To use Sherrington's term, opposing muscles were under the influence of "double reciprocal innervation." ([13] p. 94]).

By varying the relative strength of bilateral stimuli Sherrington obtained differing results. When stimulation on both sides was so arranged that the innervation of opposing muscles was equal rhythmic alternating movements in opposite phase usually developed. Variations in the strength of the opposing stimuli led to the maintenance of different attitudes of the limbs with contraction of all the muscles.

Considered in the light of these experimental results, it is likely that the rhythmic alternating movements of the hand, forearm and elbow which were observed as part of associated postural reactions in the upper limbs in some of our patients, was a manifestation of the action of two equal but antagonistic forces acting alternately. The stimulus which under this condition started and kept going these biphasic movements was the weight of the segment of the limb

itself, for they tended to appear only when the limb was unsupported. In the muscles of those joints at which displacement occurred in one direction only, concurrent innervation was unequal in antagonists and prime movers.

In our opinion the principle of double reciprocal innervation is also the physiological explanation of the tonic state of muscles at rest. In both spastic quadriplegia and hemiplegia all the muscles of the large joints accessible to inspection and palpation show hypertonicity, but tonus is developed in excess in some more than in others, hence the prevailing attitudes of the limbs and trunk at rest. Here again there is concurrent but unequal innervation of opposing muscles at different joints.

To sum up our conclusions, observations made by inspection and palpation of muscles upon the distribution of innervation in spastic hemiplegia and quadriplegia appear to show that in voluntary movements of healthy and paretic limbs and in reflex flexion movements of the upper and lower imbs there is innervation of prime movers, synergists and fixators and reciprocal relaxation of antagonists.

But in associated postural reactions of the trunk and paralysed or healthy limbs, tonic postures of the body at rest and probably reflex extension reactions of the upper and lower limbs evoked by stimulation of the axilla or groin, all muscles involved show increased contraction and are under the influence of concurrent innervation. These reactions result from unequal innervation of opposing muscle-groups; when this inequality disappears and innervation is balanced, rhythmic movements due to alternating contraction and relaxation of opposing muscles may appear.

General Summary.

In many cases of quadriplegia and hemiplegia reflex movements of the upper limbs can be elicited analogous to those which are known to be present in the lower limbs in cases of paraplegia.

In all the cases of hemiplegia recorded in this paper, with one exception, a flexion reflex of the upper limb was obtained (p. 434).

In all the cases of quadriplegia and in one case of hemiplegia an extension reflex of the upper limb was obtained. In two cases of quadriplegia it was possible, in addition, to evoke a flexion reflex of the upper limb (p. 434).

A flexion reflex of the upper limb usually comprises movements of

abduction and external rotation at the shoulder, of flexion at the elbow, wrist and finger joints. Its receptive field includes the palmar aspect of the hand and fingers, the inner side of the forearm and arm, the walls of the axilla and the upper part of the chest. The focus of the receptive field lies in the palm of the hand (p. 434). Appropriate stimuli are of a potentially painful nature, such as scratches or pinches. The response was uniphasic in hemiplegia, and biphasic in one example of quadriplegia (p. 435).

An extension reflex of the upper limb comprises movements of elevation, retraction, adduction and internal rotation at the shoulder, extension at the elbow, pronation of the forearm, flexion at the wrist and hyperextension and adduction of the fingers and thumb (figs. 2 and 3). The receptive field is co-extensive with that of the flexion reflex but its focus lies in the axillary walls (p. 432). The appropriate stimuli are similar to those of the flexion reflex (p. 428).

The spasticity of patients with quadriplegia in extension and hemiplegia presents close analogies with that of the decerebrate anthropoids described by Sherrington (p. 450).

The so-called "associated movements" are postural reactions; they were found in all the cases of quadriplegia and hemiplegia under consideration accompanying strong reflex and voluntary movements (p. 436).

In all the cases of quadriplegia and hemiplegia we have described voluntary movements and flexion reflexes of the upper and lower limbs showed the phenomenon of reciprocal inhibition of antagonists. Concurrent innervation is a feature of all postural reactions of the trunk and limbs, whether at rest or secondary to strong reflex and voluntary movements, and probably of the extension reflexes of the upper and lower limbs (p. 468).

Appendix.

Case 2.—A. A., aged 29. Following an attack of "influenzal meningitis" at the age of 1 year and 9 months the patient is said to have been weak in both lower limbs and to be troubled with involuntary movements of his head.

When 6 years old he was able to walk to school, a distance of half a mile without assistance, but he was unable to run. His progress at school was up to the average standard for his age.

He left school at 15 and went to help his brother, who carried on a grocer's business, and, although walking was difficult for him, he canvassed the houses in the neighbourhood for orders. After a year he gave up this employment to enter Beck's Optical Works in Kentish Town. He was greatly handicapped, however, by weakness and clumsiness of his movements and his tendency

to fall, and at the end of two years his employers discharged him. Since 1912 he has done no work and thinks he has got more helpless.

Since 1919, when some of the tendons of his feet were divided, he has been mostly confined to bed.

He came under our observation in December, 1920.

There is nothing worthy of note in his family history.

Summary of his Condition between December, 1920 *and December*, 1921.

He is moderately intelligent and is reconciled to his disability. He has never suffered from fits or convulsions; speech and articulation are normal. In the right ear he hears the ticking of a watch at a distance of six inches, but he is completely deaf in his left ear. His visual fields are full and his fundi are normal. His left pupil is slightly larger than his right, but they are both central, regular in outline and react well to light and on convergence. Ocular movements are full; there is no diplopia or strabismus, but nystagmoid movements are evoked by lateral fixation to either side. Sensibility to light touch and pin-prick appears to be relatively diminished on the right half of his face, except his nose. In other respects the functions of his cranial nerves are normal.

Motion: voluntary power.—All movements at the different joints can be carried out but they are decidedly weak. The left upper limb is stronger than the right; in both backward movement and adduction at the shoulder, extension at the elbow, pronation of the forearm and flexion at the wrist are stronger than the movements in the opposite directions. The thumb can be approximated to the tip of each finger, but its terminal phalanx is kept hyperextended and the movements are clumsy.

Extension of the trunk is stronger than flexion and he is unable to sit up in bed without assistance.

His right lower limb is weaker than his left; in both the weakest movements are flexion and adduction at the hip, flexion at the knee, and dorsiflexion and eversion of the foot.

The voluntary movements of his neck are not obviously impaired.

Involuntary movements.—There are occasional slight flexor movements of his lower limbs, more often on the right side. Involuntary movements of his upper limbs, apart from the associated reactions about to be described, have not been observed.

Postural reactions associated with voluntary movements.—With voluntary flexion of his left elbow against resistance the following displacements were observed: In the right upper limb adduction and backward movement at the shoulder and slight flexion at the elbow and wrist; in the trunk rotation to the right; in the lower limbs bilateral extension with downward movement of the feet and toes.

With resisted voluntary extension of the left elbow there occurred downward

movement and adduction at the right shoulder, strong extension at the elbow, pronation of the forearm, and slight extension at the wrist; the trunk rotated slightly to the left and there was bilateral extension of the lower limbs.

When the left lower limb was flexed against resistance, the movements in remote parts were slight flexion followed by extension of the right lower limb, rotation of the trunk to the left, and extension with adduction and inward rotation of both upper limbs.

Voluntary extension of the left lower limb against resistance evoked a general reaction which was approximately the same as that just described, In each of these reactions antagonistic muscles were contracted.

Gait.—With the aid of jointed mechanical supports for his lower limbs fitted to a spinal jacket, he was able to walk if helped to maintain his equilibrium.

Tone.—All the muscles acting on the large joints of the upper and lower limbs (except deltoid on the right side) and those of the trunk from the shoulder girdles downwards were spastic, but the right upper and lower limbs were more affected than those on the left side. Spasticity was developed in some muscle-groups more than in the corresponding antagonists. The more tonic muscles were the retractors, adductors, elevators and internal rotators of the shoulders, the extensors of the elbows, the pronators of the forearms, the flexors of the wrists and the extensors of the trunk and lower limbs, including the calf muscles. The fingers and toes with the exception of the adductors of the thumbs were hypotonic. The general attitude of the body at rest was one of extension with internal rotation of the upper limbs.

Sometimes after an upper limb had been placed in the position of flexion at the elbow biceps became more tonic than triceps.

The tests employed in the investigation of spasticity and the results obtained are described in the text in Chapter V.

Reflexes—The application of potentially painful stimuli to the palmar aspect of the fingers and hand, the inner and flexor aspects of the forearm or upper arm, the axillary walls or the upper part of the chest from the second to about the sixth ribs in front and behind, evoked a movement of the upper limb which was almost identical with that described in No. 1. The inner walls of the axilla were found to be the focus of the receptive field. Alteration in the situation of the stimulus within the receptive field had little effect in modifying the form of the response. By firmly pinching the tissues of the hypothenar eminence or of the forearm a quick movement of abduction at the shoulder and flexion at the elbow followed by abduction and extension could sometimes be obtained. Simultaneous stimulation on both sides elicited a bilateral biphasic response.

The abdominal reflexes were present, but diminished.

Stimulation of the sole of the foot and of the leg below the knee evoked reflex flexion of the lower limb, including upward movement of the toes. The threshold value of stimulus was lowest in the sole of the foot.

Extension at hip and knee and downward movement of the foot and toes followed appropriate stimulation of the thigh. The focus of the receptive field for the extension reflex of the lower limb was the upper part of the inner aspect of the thigh, the external genitalia and the perineum. When the nocuous stimulus applied to the thigh was intense, reflex flexion instead of reflex extension of the lower limbs was usually obtained.

The tendon reflexes of the upper and lower limbs were very brisk and knee and ankle clonus could be evoked.

Postural reaction associated with the extension reflex of the upper limb.—In the opposite upper limb there usually occurred contraction of all the muscles of the shoulder and elbow joints with some adduction at the shoulder and flexion at the elbow.

The flexors and extensors of the trunk were brought into action but more on the side of the stimulated upper limb.

The lower limbs remained rigidly extended at hips and knees, with some downward movement of the feet and toes. There was increased contraction of all the muscles, but the reaction was stronger in the ipsilateral lower limb.

Postural reaction associated with the flexion reflex of the lower limb.—The associated response appeared first in the opposite lower limb, and as in all reactions of this kind, antagonists, as well as prime movers and fixators, were brought into action. The limb remained extended at all joints. In the trunk there was strong contraction of the extensors, and to a less extent of the recti abdominales. The upper limbs became more adducted and rotated inwards at the shoulders, extended at the elbows and flexed at the wrists, with the fingers hyperextended; all the muscles at these joints, however, showed increased contraction.

Sensation.—The patient did not suffer from pain or other abnormal sensations. Sensibility to light touch was impaired on both sides below the level of the third cervical root area; the defect was greatest in the area of distribution of the fourth, fifth and sixth cervical roots, but was nowhere complete. Sensibility to pain and to heat and cold was abolished on the left side (except over the buttock) below the third cervical root area, within which painful stimuli evoked an over-reaction. On the right side sensibility to these stimuli was less affected except on the outer aspect of the upper limb and on the upper part of the chest above the second rib. Postural sensibility was grossly affected in both upper limbs, but the loss was less severe in the lower extremities.

Bladder and rectum.—There was impaired voluntary control over the bladder and rectum.

Vasomotor, sudorific and nutritional changes.—The patient was thin but there was no localized wasting, except of deltoid on the right side, which failed to respond to faradic stimuli. Gross vasomotor and sudorific disturbances were not observed.

Subsequent history.—This patient was attacked with acute lobar pneu-

monia which proved fatal. The post-mortem examination on January 5, 1922, showed incomplete, apparently traumatic, healed, transverse myelitis of the fourth and fifth cervical segments of the spinal cord.

Dr. Hubert M. Turnbull furnished the following preliminary report of the autopsy for the purposes of this article :—

There was a band of degeneration in the fourth cervical segment crossing dorsal to the canal, destroying the whole of posterior columns save two isolated dorsal areas, and dipping ventrally for a short distance into each lateral column. In the upper and central thirds of the fifth cervical segment a cross section of cord rendered trefoil by wedges of gelatinous gliosis passing inwards and ventralwards from region of posterior roots; the ventral third of the fifth cervical segment was split into some seven areas by strands of gelatinous gliosis containing minute cavities. Secondary ascending degeneration was seen in the ventral portion of the column of Goll above the fourth cervical segment; descending degeneration in dorso-lateral tracts below the fifth cervical segment. Punctate hæmorrhages were frequent in the centre of the posterior columns of the sixth thoracic segment. Dense adhesions between arachnoid and dura were present over the fourth and fifth cervical segments; considerable adhesion over the remaining cervical segments. Slight adhesion and conspicuous vascularity over the remainder of the inner surface of the dural theca. Conspicuous opaque, white thickening of the leptomeninges over the fifth and seventh thoracic segments. No thickening of leptomeninges of brain and brain-stem. Antero-posterior mobility between the bodies of third and fourth cervical vertebræ was greater than in the remainder of neck, but there was no definite evidence of previous fracture-dislocation. Cavity of the left middle ear and mastoid cell narrowed by chronic osteitis. Perforation of left drum. Mass of wax in left external auditory meatus. Scoliosis with convexity to the right in the mid-thoracic spine. Considerable lordosis. Extreme lateral compression of thorax (chicken-breast). Tapered fingers most conspicuous on the right hand. Œdema of the feet and legs. Wasting of the right thigh; great wasting of the left thigh.

Case 3.—Gunner W. J., R.F.A., aged 29. On July 15, 1916, he was wounded by pieces of shell casing. Two small wounds were found to the left of the middle line behind at the junction of his neck and shoulder and a third on the outer side of his left arm. Consciousness was retained and he believed he had full control over his bladder and rectum, but for a time his upper and lower limbs were completely paralysed. Two hours after the injury his right upper and lower limbs had begun to recover.

He was evacuated to England and on July 18 was admitted to King George's Hospital, London.

Condition on admission: He was dull and heavy, answered questions slowly and inaccurately and obviously resented examination. His temperature and pulse rate were slightly raised and he complained of headache.

The left pupil and palpebral fissure were smaller than those on the right side. There was complete paralysis of the left upper limb with the exception of slight contraction on voluntary effort of biceps and deltoid. In the right upper limb voluntary extension and flexion of the wrist and fingers could be carried out feebly.

Voluntary contraction of the rectus abdominalis was observed on the right but not on the left side.

All voluntary movements were present in the right lower limb and were fairly strong, but the left lower limb was completely paralysed.

There was complete analgesia to pin-pricks on the right side below the level corresponding to the sixth cervical segment and on the outer aspect of the left upper limb within the area supplied by the sixth cervical segment. Sensibility to heat and cold was abolished on the right side within the analgesic zone. Recognition of changes in passive position was defective in the toes of the left foot only and in the right lower limb vibrations of a large tuning-fork (C_{128}) were not appreciated. Sensibility to cotton wool was not impaired.

The tendon reflexes in both the upper and in the left lower limbs and the abdominal reflexes on both sides were abolished. The extensor form of plantar response was evoked on either side.

The left lung was dull to percussion behind and in the axilla, vocal fremitus was absent and the breath sounds were feeble. The patient was troubled with a cough and there was much expectoration.

It was reported after X-ray examination that a small fragment of metal had passed between the fifth and sixth cervical vertebræ in the region of the lateral processes on the left side; the adjacent surfaces of the vertebræ were damaged. A second fragment had travelled between the sixth and seventh cervical vertebræ.

Operation performed by Lieut.-Colonel Armour on September 1, 1916.—The spinous processes of the injured vertebræ were exposed. A piece of metal was found lying on the top of the left lamina of the seventh cervical vertebra. The laminal arch of the fifth cervical vertebra was found to be damaged and a piece of bone had been driven on to the dura. The dura was adherent to the fifth and sixth cervical vertebræ and when the membrane was opened the cord was found to be adherent to it at this level. A small piece of metal was lifted from the cord on the right side opposite the fifth cervical vertebra.

Summary of Condition up to June 26, 1917.

On September 12, 1916, the operation wound was healed, and the area of sensory loss in the left upper limb had diminished and voluntary movement of the arm had increased.

On January 29, 1917, he had full control over his bladder and rectum.

On March 13, 1917, the patient began to have involuntary extensor spasms of both upper limbs, which tended to appear as he dropped off to sleep. Voluntary power in his right upper limb was practically normal and had

improved in his left upper limb at the shoulder and elbow. His left hand when at rest was held in a position of flexion at the wrist, hyperextension at the metacarpo-phalangeal joints and flexion at the interphalangeal joints.

It was found that when the skin in the right pectoral region was scratched a reflex movement was obtained in the left upper limb, consisting of extension of the fingers, flexion of the wrist, and extension of the elbow. The reaction could be obtained by similar stimulation on the left side. When the stimulus was strong it evoked a flexor spasm in the left leg in addition to the movements in the left upper limb.

On scratching the sole of the left foot there occurred reflex flexion of the left lower limb with upward movement of the foot and toes and extension of the right knee with upward movement of the foot and toes. On scratching the sole of the right foot reflex flexion of the right lower limb and extension of the left lower limb with downward movement of the foot and toes were obtained.

Later observations of the extension reflex of the left upper limb showed that the reaction included adduction and internal rotation of the shoulder and adduction of the thumb with flexion of its terminal phalanx. There was also adduction of all the fingers. The receptive field of the reflex response, which was strictly confined to the left upper limb, was bilateral and included the palmar aspect of the fingers and hands, the flexor and inner aspects of the forearms, the inner aspects of the upper arms, the walls of the axillæ and the upper part of the chest in front and behind between the second and fifth ribs.

Recovery of voluntary power continued and in May, 1917, he was able to flex and extend his fingers, thumb and wrist on the left side and to pronate and supinate his left forearm. Voluntary movements at the elbow and shoulder had improved. In the left lower limb all movements at hip and knee were good and he was able slightly to move his toes up and down, but movements at the ankle were absent.

Case 4.—A. B. C., aged 19. Jewel-case maker. Came under observation on February 14, 1921.

Three years ago he suffered from an illness of three weeks duration and diagnosed as "influenza." When he began to get about he noticed weakness and stiffness of both lower limbs on the right more than the left side. The disability slowly increased, and a year later his left upper limb became affected in the same way. At this time he began to suffer from sharp stabbing pains in his lower limbs, chiefly in the anterior aspects of the thighs and behind the knees. The pain was intermittent, and was aggravated by cold and wet weather and by fatigue. There were also occasionally pricking pains in his fingers and arms, which were associated with involuntary abduction movements of his upper limbs, and any object that he happened to be holding in his hands at the time was thrown to the ground.

During the past year his disability has been stationary.

Previous history.—He had measles as a child, and pneumonia at the age

of seven. He was liable to attacks of bronchitis each winter up to the age of fourteen years.

His family history reveals nothing of interest in connection with the case.

Summary of Condition at the Present Time.

He is bright, intelligent, and gives a good account of himself; he sleeps well, and appears to suffer little discomfort. Speech and articulation are normal. He has never suffered from fits or convulsions. Occasionally frontal headaches trouble him, but they are never associated with vomiting. Hearing and vision are good on both sides, and his discs and fundi are normal. Apart from nystagmoid movements on extreme lateral fixation of his eyes to the right or left the functions of his cranial nerves are normal.

Motion: voluntary power.—He is able to perform all voluntary movements with his trunk and limbs, but they are weak; his left upper limb is more affected than his right.

In the upper limbs voluntary movements of extension are weaker than those of flexion, and the opposite is the case in the lower limbs.

Involuntary movements.—Involuntary movements of extension of his lower limbs sometimes occur in the morning when he wakes. As previously mentioned, involuntary quick extension of an upper limb is liable to follow the presence of sharp, pricking pains in the fingers and arm, to which he is sometimes liable. Knee-clonus, unilateral or bilateral, is frequently observed when the patient lies in bed; it occurs particularly when the patient is excited for any reason.

In the morning when he yawns on waking, his upper limbs become extended above his head, and this posture tends to persist for some time after the act of yawning has ceased. As part of this automatic act the trunk and lower limbs are rigidly extended.

Tone.—All the muscles of his limbs, trunk and shoulder-girdle, including trapezius on both sides, are spastic. The degree of spasticity, however, is not excessive, but is unequal in its distribution; thus the lower limbs are more affected than the upper limbs, and the left more than the right upper limb.

The muscles of the shoulder-girdle are all affected equally, but the flexors of the elbows, the pronators of the forearm, the flexors of the wrist, and the extensors of the trunk and lower limbs are more hypertonic than their corresponding opponents.

Nutrition.—The patient is thin, but there is no local wasting in any part of the body.

Gait.—He is able to walk stiffly without support. In walking his right knee is, as a rule, slightly bent, and he walks on the balls of the toes of his right foot. His left lower limb is rotated inwards.

Reflexes.—All the tendon jerks in the upper and lower limbs are much increased, and are equal on the two sides.

There is bilateral knee and ankle clonus. The clonus, when present in one lower limb, can be immediately arrested by evoking either a flexion reflex

in the same or opposite lower limb. In the latter case the flexion reflex is associated with increased tone in all the muscles of the crossed lower limb, but mostly in the extensors.

The abdominal reflexes are present in all segments, but are brisker on the right than on the left side.

Reflex flexion of the lower limb can be evoked by stimulation of the sole of the foot or of any part of the leg below the knee. The outer part of the sole of the foot forms the focus of the receptive field for the reflex. Stimulation of the thigh of mild intensity usually excites extension of the limb on the same side. The focus of the receptive field for the extension reflex lies in the region of the groin, perineum and external genitalia.

Both reflex flexion and reflex extension are accompanied by extension of the crossed lower limb with downward movement of the foot and toes, although tone in all the muscles of the limb is increased.

Reactions evoked by stimulation of the upper limb or upper part of the trunk.—Gentle stimulation of the palm of the hand, the inner aspect of the upper limb, the walls of the axilla or the upper part of the chest, in front or behind, evokes the following reaction, which varies much in detail, according to the locality of the stimulus within the receptive field. The general response consisted of a quick movement of elevation, adduction with external rotation at the shoulder, flexion at the elbow, and extension of the wrist and fingers. There is usually a slight delay between the application of the stimulus and the appearance of the reaction and relaxation, after the primary movement has occurred, is relatively slow. The response is most easily excited by stimulation of the inner aspect of the upper arm and especially of the walls of the axilla. In this region a gentle moving contact with the tip of the finger, and sometimes a mere touch, is sufficient to excite a reaction.

There is great adaptability in the form of the response to the locality of the stimulus. Thus when the reaction is evoked from the palmar aspect of the hand or fingers, the main part of the reaction consists of extension of the fingers and hand, adduction of the thumb and flexion of the elbow. When the stimulus is applied near the elbow, abduction and internal rotation at the shoulder with flexion at the elbow are prominent. Extension at the elbow, however, is invariably obtained from stimulation of the skin behind this joint. When the stimulus is applied to the inner aspect of the upper arm or to the axilla, two main forms of response are obtained. Adduction and external rotation at the shoulder form the main part of the movement with stimulation of the front part of this region, and abduction and internal rotation when the point stimulated lies within the posterior part of this area.

The outer aspect of the upper limb is also responsive but to a less extent than the inner aspect. Scratching the skin over the deltoid, for example, evokes adduction, strong elevation and external rotation of the shoulder.

Reactions of the upper limb on the same side are also evoked by stimulation of the trunk. Pinching the skin over the upper abdominal wall or lower ribs in front evokes a movement in which the trunk is bent towards the stimulated

side. When the point stimulated, however, is in the neighbourhood of the lower border of the fold formed by pectoralis major, this lateral movement of the trunk tends to be accompanied by a response in the upper limb on the same side consisting of adduction and external rotation at the shoulder, slight flexion at the elbow and extension of the wrist. When the stimulus is applied to the supraclavicular region the same reaction in the upper limb is obtained but elevation of the shoulder is prominent. Stimulation of the skin over the scapula evokes retraction of the upper limb and when the stimulus is applied near the inferior angle of the scapula there occurs, in addition, downward movement of the shoulder, flexion at the elbow, and lateral displacement of the head to the stimulated side. When the situation of the stimulus lies over the upper fold of trapezius near the neck the response consists mainly of elevation and forward movement of the shoulder.

By scratching the skin in the middle line over the upper thoracic spinous processes a backward twitch of the head is evoked along with a bilateral quick movement of elevation of the shoulders. By moving the stimulus downwards to the inter-scapular region in the middle line, the elevation of the shoulders and backward movement of the head is now accompanied by quick backward movement of the shoulders followed by forward movement of these parts.

Motor phenomena in association with reflex flexion of the lower limbs.—Associated reactions are not observed with nociceptive reflex movements of the upper limb but with reflex flexion of the lower limb the following general response is usually obtained. The crossed lower limb is strongly extended at hip and knee and the foot and toes move down. All the muscles of the limb, however, are brought into action, as also are the muscles of the trunk; in the upper extremities movement as a rule does not occur, but all the muscles acting on the shoulder and elbow-joints on both sides show increased tone.

Motor phenomena associated with strong voluntary movements.—When the patient makes strong efforts to extend his left elbow against resistance, slight abduction is observed at the right shoulder, flexion at the elbow, supination of the forearm, and extension of the wrist; the trunk and lower limbs are extended. All the muscles are brought into action, movement at different joints being determined by stronger contraction of some muscles over their corresponding opponents.

With voluntary flexion of the left elbow against resistance, all the large muscles of the right upper limb become more tonic, but very little movement occurs; the arm is held a little abducted and internally rotated at the shoulder, the elbow is held in a position of flexion or extension, the posture being determined by the position of the joint before the response is evoked; the forearm is supinated to a slight extent and the hand is extended; the trunk is often slightly bent forward, and the lower limbs are strongly extended.

Similar reactions are evoked in association with voluntary flexion or extension of the right elbow against resistance.

Strong voluntary flexion or extension of one knee against resistance also evokes a general response in the rest of the body. The knee of the opposite

lower limb usually becomes slightly flexed, although all the muscles are tonically contracted as are also those of the trunk, shoulder-girdle and elbow-joints.

Sensation.—Sensibility is unimpaired, except for defective recognition of vibrations of a large tuning-fork in the lower limbs and of changes in passive position in the toes.

Bladder and rectum.—There is definite frequency of micturition, sometimes associated with hesitancy. Aperients have to be given daily to keep his bowels open.

Vasomotor and nutritional functions are normal.

X-ray examination of the spine shows no abnormality.

Case 6.—L. S., a man, aged 27, was admitted into St. Thomas's Hospital on August 26, 1921.

During the previous week he had experienced a certain amount of abdominal discomfort. On the day of admission he complained in the afternoon of some stiffness of his neck, but continued at his clerical work. After tea he felt unwell and fainted and a few minutes later began to lose power in his right hand and arm and then in his right leg. Twenty minutes later he was deeply unconscious, vomited two or three times, and was brought into hospital.

On examination in bed he was found in a comatose condition with a full and regular pulse and deep regular respiratory movements.

The left limbs presented some resistance to passive movements, but the right arm and leg were completely flaccid. The reflexes on the right side were of the usual hemiplegic character, those on the left being normal.

Three days later the patient was semi-conscious, but more restless and irritable under examination. Cerebrospinal fluid withdrawn by lumbar puncture was under increased pressure and blood-stained.

The Wassermann reaction in the blood and cerebrospinal fluid was negative. The cerebro-spinal fluid contained many blood-cells and a slight excess of lymphocytes. Cultures of the fluid proved sterile.

During the next week or two the patient gradually recovered consciousness, but was found to suffer from a considerable degree of motor aphasia and also from severe headache, which gradually grew less intense.

The cerebrospinal fluid was examined on several occasions, and showed a progressively decreasing amount of blood as time went on. Voluntary power together with an increasing degree of rigidity developed in his right limbs, and by the middle of October he presented a typical picture of spastic right hemiplegia.

Posture.—The right upper limb was held in a position of adduction and slight internal rotation at the shoulder, flexion at the elbow, semi-pronation and flexion at the wrist and flexion at all finger-joints: the right lower limb was in a position of general extension at all joints.

Tone.—There appeared to be a general increase of tone throughout the muscles of the right upper limb with distinctly more tone in the adductors of the

shoulder, flexors of the elbow, and pronators and flexors of the wrist and fingers. In the right lower limb the amount of tone was not so pronounced but was relatively greater in the extensors than in the flexors.

Reflexes.—All tendon-jerks in the right upper and lower limbs were greatly exaggerated. The right abdominal reflexes were less brisk than those on the left side. Stimulation of the right foot produced a general flexor response of the right limb accompanied by an upward movement of the big toe.

A nocuous stimulus, such as the scratch of a pin or a firm pinch applied to the palm of the right hand or to any part of the inner aspect of the right arm and forearm or to the anterior wall of the axilla, evoked a contraction in the right deltoid, infraspinatus, biceps and flexors of the wrist. The contraction of these muscles produced slight abduction and external rotation at the shoulder, flexion and supination at the elbow and slight flexion at the wrist, and was sometimes associated with extension of the thumb. It was noticeable that the contraction of the above-mentioned muscles was accompanied by relaxation of their antagonists.

This reflex was not strong enough to produce great displacement of the segments of the limb and it was not associated with any definite movements in the trunk or other limbs.

Associated postural reactions.— (1) Reflex flexion of the right lower limb, produced by stimulating the sole of the foot, was associated with slight extension at the left hip and knee, with contraction of the right abdominal wall, and with a movement of the right upper limb, which consisted of an increase of internal rotation at the shoulder, slightly more pronation and flexion at the elbow, and some flexion of the fingers.

(2) When strong voluntary movements against resistance were performed by the right upper limb, such as flexion of the fingers, flexion and extension of the elbow, and adduction and abduction of the shoulder, a number of associated movements took place in other parts of the body. There was general bilateral contraction of the trunk and neck muscles, including both trapezii and all the muscles of the abdominal wall. In the left arm there was observed abduction and internal rotation at the shoulder, flexion at the elbow, pronation of the forearm, with strong flexion of the wrist and fingers. In the lower limbs there was a slight definite movement of flexion on the right side and one of extension, chiefly at the proximal joints, on the left side.

(3) Stimulation of the sole of the left foot produced a downward movement of the left big toe, with slight dorsiflexion of the ankle, and slight flexion of the knee. This was sometimes associated with adduction and retraction of the right shoulder.

(4) Powerful voluntary movements against resistance carried out by the left arm or either leg were always associated with certain movements of the right upper extremity, and these consisted of abduction and internal rotation at the shoulder, flexion at the elbow, slight pronation of the forearm, and flexion of the wrist and fingers. If the effort was sufficiently powerful, contraction took place in the trapezii of both sides as well as in other trunk

muscles, and these movements were associated with slight flexion of the right lower limb, and slight extension of the left.

It was noticeable that in these associated movements the contraction of the prime movers was accompanied by a less powerful contraction of their antagonists.

Case 7.—A. E., a man, aged 28, was admitted to St. Thomas's Hospital, on October 13, 1921, complaining of pain in the left side of his head, general weakness and attacks of transient blurring of vision of a few months duration. There was a history of occasional nocturnal incontinuence of urine and of defective memory and attention.

On examination there was found gross papillœdema in both eyes, slight weakness of the right side of the face and of the right grasp associated with very slight impairment of postural sensibility in the right hand. The reflexes were generally normal and what changes were observed from time to time were variable and indefinite.

On November 2, 1921, Mr. Sargent explored the left fronto-temporal region of the brain and found a deep-seated tumour in the substance of the temporal lobe about one and a half inches behind the Sylvian point. No attempt was made to remove the growth, which had the appearance of an infiltrating glioma.

A post-operation right hemiplegia gradually cleared up, but on November 14 the patient became semi-comatose and had general convulsions chiefly affecting the right arm and leg. These recurred at intervals during the next few days and finally left him with a complete flaccid right hemiplegia.

On November 15 the right upper limb was completely paralysed and toneless. No tendon-jerks could be obtained either at the elbow or the wrist. Pinching or scratching the inner surface of the hand or forearm evoked a movement of abduction and external rotation at the shoulder, the movement being produced by contraction of the deltoid and infraspinatus muscles.

On November 22, a similar stimulus provoked in addition a slight contraction of the biceps with a movement of flexion at the elbow and supination of the forearm.

A week later these reflex movements were obtained with greater ease, and in addition to the movements described above there was observed slight flexion at the wrist. The whole limb was still completely lacking in voluntary power, in tone and in tendon-jerks.

If the patient carried out powerful voluntary movements of the left arm against resistance, they were accompanied by slight flexion of the right elbow and perhaps slight pronation of the right forearm.

At the end of December the right upper limb exhibited a slight amount of muscular tone and the tendon jerks could be elicited. Slight voluntary movements were possible at the shoulder and elbow, and the flexion reflex movements included well-marked flexion at the wrist.

Case 8.—W. S., a man aged 58, who was admitted into St. Thomas's Hospital on March 26, 1921.

He gave a history of fairly good health until the last two years, during which he has suffered from sharp intermittent pains in the lower extremities, and from a certain amount of difficulty in walking steadily. More recently, according to his wife, his memory and general intelligence appeared to be failing.

On March 25, 1921, after a restless night, he was discovered in the morning unable to speak, and with weakness of his left limbs.

On admission to the hospital he looked ill and somewhat emaciated. He lay quietly in bed, and could answer questions slowly, but with defective articulation. There was complete incontinence of both sphincters. The head and eyes were turned somewhat to the right, and the pupils were very small and immobile; both upper lids showed a certain amount of drooping and the patient appeared unable to turn his eyes to the left. The left side of the face was weak when compared to the right. The left upper extremity was also very weak, the muscles of the shoulder and elbow being somewhat spastic, those of the wrist and hand flaccid. The left lower extremity was weaker than the right, especially in regard to all flexor movements. There was some impairment of sensibility to both deep and superficial painful stimuli in both lower extremities.

Reflexes.—The arm-jerks were brisk and equal on the two sides. The abdominal reflexes were absent on the left side. Both knee and ankle jerks were not obtained, and a plantar response on the left side was of the extensor type, that on the right side flexor.

Cardio-vascular and respiratory systems presented no marked abnormality.

The tongue was heavily coated, and the abdomen apparently normal.

On March 27 a lumbar puncture was performed and clear cerebrospinal fluid removed. The examination of this fluid showed that it gave a positive Wassermann reaction, and contained a large excess of lymphocytes. There were some increase in the albumin content.

Although treated with intramuscular injections of mercury and two intravenous injections of N.A.B., the patient showed little sign of improvement, became more incoherent and drowsy, and died on April 13.

While under observation it was noticed that stimulation of the inner aspect of the left arm and of the palm of the left hand by means of pricking or a scratch evoked a reflex movement of the left upper limb, which consisted of internal rotation and adduction at the shoulder, extreme extension of the elbow, hyperpronation of the forearm, with slight flexion of the wrist and fingers. This "extension reflex" had a somewhat sudden onset after a latent period in response to appropriate stimulation.

The post-mortem examination revealed an area of softening, involving the posterior part of the right lenticular nucleus and the right internal capsule. There was general thickening of the pia mater, and sclerotic changes in the cerebral vessels. The spinal cord showed chronic lepto-meningitis involving the posterior surface with atrophy of the posterior columns and the posterior roots.

There were œdema and congestion of the lungs, chronic interstitial nephritis, atheroma of the aorta, and sclerotic changes of the cardiac valves.

REFERENCES.

[1] Babinski, "Du phénomène des orteils et de sa valeur sémiologique," *Semaine Médicale*, 1898, **18**, 321.

[2] Beevor, Charles. "The Croonian Lectures on Muscular Movements and their Representation in the Central Nervous System," London, 1904, 54.

[3] Brown, Graham. "On the Occurrence of a Plastic Flexor Tone in the Monkey," *Journ. of Physiol.*, 1915, **49**, 180,

[4] Head, Henry, and Riddoch, George. "The Automatic Bladder, Excessive Sweating and some other Reflex Conditions in Gross Injuries of the Spinal Cord," *Brain*, 1917, **40**, 217.

[5] Kelling, Georg. "Untersuchungen über die Spannungszustände der Bauchwand," *Zeitschr. f. Biol.*, 1903, Bd. **44**, S. 161.

[6] Riddoch, George. "The Reflex Functions of the Completely Divided Spinal Cord in Man," *Brain*, 1917, **40**, 264.

[7] *Idem.* "The Clinical Picture of Complete Transverse Division of the Spinal Cord," *Medical Science*, 1921, **5**, 49.

[8] Sherrington, C. S. "The Integrative Action of the Nervous System," London, 1906.

[9] *Idem.* "Cataleptoid Reflexes in the Monkey," *Proc. Roy. Soc.*, 1897, **60**, 412.

[10] *Idem.* "Flexion-Reflex of the Limb, Crossed Extension Reflex and Reflex Stepping and Standing," *Journ. of Physiol.*, 1910, **40**, 105.

[11] *Idem.* "The Postural Activity of Muscle and Nerve," *Brain*, 1915, **38**, 212.

[12] *Idem.* "Decerebrate Rigidity and Reflex Co-ordination of Movements," *Journ. of Physiol.*, 1898, **22**, 319.

[13] *Idem.* "Reciprocal Innervation." Seventeenth International Congress of Medicine, London, 1913, Section II, Physiology, part i, 85.

[14] Tilney and Riley. "The Form and Functions of the Central Nervous System." London, 1921, 154.

[15] Viets, Henry. "Relation of the Form of the Knee-jerk and Patellar Clonus to Muscle Tonus," *Brain*, 1920, **43**, 269.

[16] Walshe, F. M. R. "The Physiological Significance of the Reflex Phenomena in Spastic Paralysis of the Lower Limbs," *Brain*, 1914, **37**, 269.

[17] *Idem.* "On the Genesis and Physiological Significance of Spasticity and other Disorders of Motor Innervation," *Brain*, 1919, **42**, 16.

[18] Wilson, S. A. Kinnier.—"On Decerebrate Rigidity in Man and the Occurrence of Tonic Fits," *Brain*, 1920, **43**, 220.

DYSSYNERGIA CEREBELLARIS MYOCLONICA—PRIMARY ATROPHY OF THE DENTATE SYSTEM: A CONTRIBUTION TO THE PATHOLOGY AND SYMPTOMATOLOGY OF THE CEREBELLUM.[1]

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CHAPTER I.—INTRODUCTION.

SOME years ago, under the title "Dyssynergia Cerebellaris Progressiva, or Chronic Progressive Cerebellar Tremor" [10] I directed attention to a peculiar disorder of motility, which I regarded as a definite clinical type of nervous disease.

This affection was characterized by generalized intention tremors, which began as a local manifestation and gradually extended to other parts of the voluntary muscular system. The extremities, and more especially the arms, showed the greatest degree of involvement.

The coarse ataxic-tremor disturbance which characterized the dis-

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order was only present when the muscles were in action, and ceased entirely during relaxation and rest. It was, therefore, volitional in character and of the "intention tremor type." Another peculiar feature was the severe involvement of the finer co-ordinated movements of the extremities, while the coarser and more massive movements underlying station and locomotion were but little affected.

When this tremor disturbance was subjected to careful analysis there was found associated with it a disorder of muscle tone and of the ability to measure direct and associate muscular movements, the clinical manifestations of which were dyssynergia, dysmetria, adiadokokinesis, hypotonia and asthenia. All of these symptoms, including the volitional tremor, which was an extreme expression of the underlying disturbance of muscle tone and synergy, showed the existence of a fundamental disorder of cerebellar function.

I, therefore, regarded the affection with its progressive tendency, chronic course and well-defined cerebellar symptomatology as "an organic disease caused by degeneration of certain special structures of the cerebellar mechanism, which were concerned in the regulation of the tonus and synergies of muscles."

Since my original publication, I have had occasion to observe another group of cases which combined the symptomatology of dyssynergia cerebellaris progressiva and myoclonus-epilepsy. There was the progressive dyssynergia characteristic of a cerebellar disorder, in association with epilepsy and myoclonus. Here, as in the group of cases uncomplicated by myoclonus-epilepsy, the movements of the extremities showed the greatest degree of disturbance.

This form of dyssynergia, which I would term dyssynergia cerebellaris myoclonica, I have also observed in association with Friedreich's ataxia, a combined cerebello-spinal involvement, which is not infrequent in the history of cerebellar system disease. In one of these cases careful pathological investigations were made which throw considerable light on the origin and anatomical basis of the cerebellar portion of the symptomatology. On the basis of these studies I would refer the progressive dyssynergia to a primary atrophy of the efferent dentate system of the cerebellum, and would regard this system as the essential neural mechanism underlying the production of the cerebellar or intention tremor.

In conclusion, I will state my views on the relation of the cerebellum to the static or posture system of motility, and the bearing which this conception has on the interpretation of cerebellar function.

Chapter II.—Dyssynergia Cerebellaris Progressiva Myoclonica: a Form of Progressive Cerebellar Tremor associated with Myoclonus-Epilepsy.

Critical Report of Four Cases.

Case 1.

Summary.—Myoclonus-epilepsy began at the age of 17, and was followed five years later by symptoms of dyssynergia cerebellaris progressiva, viz., generalized intention tremors and scanning speech, associated with dysmetria, dysdiadokokinesis, hypotonia and asthenia.

The patient, a man aged 30, single and a telegraphist by occupation, was referred to the Neurological Institute, in May, 1918, and was admitted to the service of Dr. Pearce Bailey. I am indebted to Dr. Bailey for the privilege of observing and reporting the case.

Family history.—His father was a periodical drinker and two of the father's uncles were alcoholic. An aunt on the father's side was subject to fainting attacks of rather doubtful nature and which were not called epileptic by the family. Their character could not be determined with certainty. His mother is living and in good health. Parents were not related. One sister is alive and well. A brother died in early life. No other member of the family, so far as he knows, has had any disease similar to the one from which he is suffering.

The previous history shows nothing unusual. Childhood was normal, he did good work at school and was quite up to the average in his mental development. He had measles and whooping-cough in early life. There was no history of trauma, venereal disease or of alcoholic excesses.

Onset.—The first symptoms of his disease appeared at the age of 17. At this time he had an epileptic seizure which was characterized by clonic muscular spasms of the right arm and leg, which developed while wrestling and lasted only a few minutes. He had two such attacks, neither of which were associated with complete unconsciousness but in both he became dizzy and fell to the ground. After these initial seizures the subsequent attacks were always accompanied by complete loss of consciousness. They were nearly always preceded by a motor aura, consisting of a progressive spasm of the right arm and leg, which terminated in unconsciousness, and was followed by a general convulsion. The tongue was often bitten, and there were occasional incontinence of urine and the other manifestations of a major epileptic seizure. These grand-mal attacks were comparatively infrequent, occurring only three or four times a year, and at one time there was a free interval of nearly three years.

When he was 19 years of age, that is, two years after the first epileptic seizure, the myoclonic jerks made their appearance. These were usually worse in the early morning hours and gradually increased in extent and intensity. He states that they were usually diminished by the use of alcohol, and the

patient sometimes took this means of relieving their severity. The myoclonus first appeared in the hands and arms and gradually extended to the trunk and lower extremities. Subsequently, the myoclonus became so severe that he was occasionally thrown to the ground and on one of these occasions he broke his nose.

Five years after the onset of the myoclonus-epilepsy the intention tremor made its appearance in the arms. This gradually increased in severity and later the speech became affected, and the legs showed evidences of involvement.

There has been no headache, no diplopia and he states no special impairment of memory. Preceding the epileptic attacks there is a whirring sensation in the head but otherwise no vertiginous seizures. The myoclonus is increased by mental stress and worry, and varies in its intensity from day to day. He has good days when the involuntary jerks and starts are in abeyance and other days when they are very severe.

Physical examination.—Shows a well-developed muscular man. The speech is slow and scanning in character and of the cerebellar dysarthric and explosive type. There is a well-marked, typical intention tremor of both upper extremities, similar in all respects to that observed in multiple sclerosis. In addition there is also a typical intention tremor of both lower extremities. Associated with the intention tremor are evidences of dysmetria and dysdiadokokinesis.

There is no loss of the gross motor power of arms or legs and no muscular atrophy. There is, however, in the arms, some diminution of the power to maintain fixed muscular contractions and postures except for short periods of time, due to tremor and asthenia. For example, when he extends the arm from the body in abduction, and attempts to hold it in this position, there ensues a coarse ataxic tremor and after a brief effort the arm is dropped to the side, in this respect simulating the behaviour of the intention tremor in multiple sclerosis. There is difficulty in maintaining a fixed posture of the upper extremities except for short periods of time. There is also a moderate degree of hypotonia in both upper and lower extremities. The Stewart-Holmes sign of cerebellar hypotonia is however not demonstrable.

The tendon reflexes of both upper and lower extremities are present, active and equal on the two sides. There is no ankle clonus, no patellar clonus and the plantar reflex is of the normal flexor type (no Babinski response). The cremasteric and abdominal reflexes are present and equal on the two sides. There is no history of pain or of paræsthesias. All sensations both superficial and deep are normal.

The pupils are equal and react to light and accommodation. The ocular excursions are normal and there is no nystagmus. Fundi negative. The innervation of the face, tongue, soft palate and muscles of mastication are also normal. Save for occasional myoclonic jerks there is no Rombergism and the station is normal. The spine is normal in contour and in movement.

During the course of the examination there are frequent myoclonic starts or jerks in both upper and lower extremities and trunk. This tendency is

increased or may be induced by voluntary movement, and this together with the intention tremor produces a very grave disorder of muscular co-ordination.

When lying on the back patient is relaxed and perfectly quiet, but any attempt at movement is likely to initiate myoclonic twitchings of the face, extremities or trunk. This tendency is increased by talking, or indeed by any form of volitional movement. In the recumbent position he can elevate both the upper and lower extremities, and hold them in this position with closed eyes without developing ataxic movements. The myoclonic jerks also occur spontaneously apparently without the intervention of volitional effort.

He is able to stand and walk, but does so with great caution and some uncertainty as if anticipating a myoclonic jerk. The intention tremors of the arms and legs occur with all volitional movements and present all the typical characteristics of this form of tremor. It is readily distinguished from myoclonic jerks although frequently accompanied by them. Associated with the intention tremor are typical dysmetria and dysdiadokokinesis. The gait and station, while showing some uncertainty, are not markedly titubating in character and the evidences of cerebellar disorder affect deeply the higher forms of motility, viz., speech and the volitional movements of the extremities (appendicular dyssynergia).

The heart and lungs are normal. Abdominal palpation is negative. There is no demonstrable enlargement of the hepatic area, and no evidences of abnormal pigmentation of the skin or cornea. Urine normal; Wassermann test of blood negative.

Comment.—This patient presents in addition to the symptoms of myoclonus-epilepsy a chronic progressive dyssynergia of cerebellar origin. With the scanning speech and intention tremors of both upper and lower extremities there are the associated manifestations of cerebellar disease, viz., dysmetria dysdiadokokinesis, hypotonia and asthenia. With the exception of epilepsy, myoclonus and dyssynergia there are no other evidences of organic disease of the nervous system. It is of interest to note that the evidences of dyssynergia are appendicular rather than truncal in distribution and that higher types of movement are chiefly affected. There was no familial history of either myoclonus-epilepsy or cerebellar disease.

Case 2.

Summary.—A girl, aged 19; onset of myoclonus-epilepsy, at the age of 7. Associated with this is a progressive dyssynergia of cerebellar origin, affecting more especially the speech and extremities.

This patient is 19 years of age, and single. She was admitted to the Craig Colony for Epileptics on January 14, 1918, and is still under observation in that institution, where I saw her in consultation with Dr. Doolittle of the Colony Staff.

Family history.—Father has been excessive in the use of alcohol. Her mother is in good health. Parents are not related. She has three brothers

and two sisters, all in good health. There is no family history of any similar nervous disorder.

Personal history.—The patient was born at full term following a labour lasting forty-eight hours. The delivery was said to have been natural. Among possible pre-natal influences may be mentioned her father's alcoholism and mother's worry over family affairs. There was no paralysis or spasm immediately after birth. Her mind developed naturally during infancy and childhood, and she was considered very bright in school. She had whooping-cough at 10 years, measles at 13 years, and diphtheria at 14.

Present illness.—At the age of 7 it was first noticed that her hands would shake and jerk while feeding herself or trying to pick up things. This tendency to myoclonus continued and gradually increased in extent and severity, finally involving, in addition to the arms, the legs, trunk, and even the facial musculature.

Three years later, when she was 10 years old, she had her first epileptic seizure. These attacks recurred every three or four months. They were diurnal and of the grand-mal type. At the present time she averages about six grand-mal seizures a year. They are of typical character, and are frequently associated with biting of the tongue and incontinence of urine. It was difficult to obtain any very definite statement as to the time of the appearance of the intention tremor, because of the existence and severe nature of the myoclonic phenomena which tended to mask the other motor disorder.

For the last five years, however, a coarse ataxic tremor of the arms and legs has been present on volitional movements and the speech has been jerky, irregular, explosive. These symptoms have grown gradually worse. It is more than probable that the characteristic symptoms of a cerebellar disorder were present before the fourteenth year, and were overshadowed by the myoclonia.

Physical examination (February, 1918).—During the course of the examination, while patient is seated in a chair, there are frequent sudden myoclonic jerks or starts, which may appear spontaneously, but usually precede or follow some volitional effort. These may appear in the arms, trunk, legs, or face; as a rule, there is a single coarse jerk or start, and then an interval—occasionally a small series of myoclonic starts occur in rapid succession. These movements are brusque, sudden, and involve synergic groups of muscles and produce a considerable locomotor effect. On arising from a sitting posture there is at first a slight unsteadiness and an effort to restore the equilibrium by standing upon a broad base with balancing. While walking, the harmony of movement is frequently broken by coarse myoclonus jerks which throw her to one side, causing a loss of balance.

These coarse myoclonus jerks appear in the trunk and arms and legs. They also occur while sitting quietly in a chair, producing sudden brusque jerks of the trunk or extremities. A slight flicker of myoclonic character is occasionally observed in the face. The myoclonus is very apt to occur just before, apparently initiated by the response to a voluntary movement, such as lifting up the arm, placing finger on the tip of the nose, or a movement of a

leg. At certain times, and on certain days, they are much more severe than at others, and in the morning they occasionally cause her to toss about in the bed so that she almost falls out. This patient's myoclonic movements sometimes continue during sleep and disturb her rest to a considerable extent. The abdominal muscles are also occasionally the seat of spasm. The myoclonic movements of the extremities are often symmetrical, both sides being simultaneously involved. The difficulties of gait and station are very much increased by the tendency to myoclonus, as all of the usual muscular activities of standing and walking in themselves produce myoclonus, and are the cause of constant uncertainty and inco-ordination.

She is able to stand alone, and there is no Rombergism. She walks on a rather broad base, lifting the foot too high with jerky, irregular ataxic-like movements of the legs (dysmetria). The attitude of the trunk is also somewhat oscillating, showing a slight tendency to balancing. The spine is straight, and there is no scoliosis.

A cerebellar or intention tremor is present in both upper extremities, more particularly on the left side. It is of the same character as the intention tremor of multiple sclerosis and is brought out conspicuously by the finger-nose test. On the performance of this test there is the coarse atactiform shaking which increases in intensity as the finger approaches the tip of the nose. And there is the quick jerking away movement of the fingers from the tip of the nose, as in true intention tremor. Well marked adiadokokinesis and evidences of dysmetria are also present on both sides.

In the recumbent posture there is also dysmetria in the heel-knee test, and volitional movements are irregular, jerky and of the cerebellar ataxic type. The movements during these tests are frequently interrupted by myoclonic jerks. There is no noteworthy hypotonia of the extremities and the Stewart-Holmes sign of hypotonia is absent.

The tendon reflexes of the arms (supinator, biceps and triceps jerks) are present and not exaggerated. The knee-jerks are active and equal on the two sides. Both Achilles jerks are present and equal. There is no clonus. The abdominal reflexes are present and equal. The plantar reflexes are also present and of the normal flexor type (no Babinski).

The pupillary reflexes are normal. The ocular excursions are normal and there is no nystagmus. Fundi normal. The innervation of the face, tongue, soft palate and muscles of mastication are normal.

The speech shows well-marked dysarthria of the cerebellar type. It is jerky, explosive and at times scanning.

There is no Romberg symptom and no ataxia of station develops on closure of the eyes. In the recumbent posture both the upper and lower extremities may be elevated and held in this position with closed eyes without the development of ataxic movements.

Sensation.—The superficial sensibility is normal. The sense of touch, pain and temperature are well preserved. The deep sensibility is also normal and there is no disorder of the sense of posture.

There is no disturbance of the sphincter control of the bladder or rectum.

The intelligence shows some general reduction and retardation. There is loss of memory and slowness of thought and action. Judgment is poor. In the Binet-Simon test she is rated at 9 years.

The heart and lungs are normal. The urine is normal. Wassermann test of the blood is negative. There are no evidences of abnormal pigmentary deposits on the skin or at the sclero-corneal junction.

Comment.—With the exception of myoclonus-epilepsy and progressive dyssynergia there are no other evidences of organic disease of the nervous system. The chief disorder of cerebellar function is that affecting articulation and the movements of the extremities. The gait and general equilibrium also show some disorder of cerebellar function, which is accentuated by the severe type of myoclonia which is present.

In this case the dyssynergia while preponderantly appendicular in distribution also involves to some extent the trunkal musculature. As in the previous case, there was no history of the familial occurrence of either myoclonus-epilepsy or cerebellar disease.

Case 3.

Summary.—Myoclonus-epilepsy of twenty-six years' duration. Associated with symptoms of dyssynergia cerebellaris progressiva, viz., cerebellar dysarthria, intention tremor, dysmetria and adiadokokinesis, affecting chiefly the volitional movements of the extremities.

This patient was a man, aged 38, single, an inmate of Craig Colony for Epileptics, where I saw him in consultation with Dr. Shaw of the Colony staff.

Family history.—Parents are living and in good health, patient is an only child of his fraternity. He has one half sister (mother's daughter by first marriage), aged 42, who is married and in good health. Careful inquiry by letters and interviews failed to reveal any history of epilepsy or other nervous disorder among the relatives.

Personal history.—The history of the birth, of infancy and of childhood were negative up to the age of 12 years. He had whooping-cough at 8, and measles at 9. There were no untoward symptoms during these infections, which would indicate involvement of the central nervous system. School progress was about the average, and at the age of 16 he left school to work on a farm. He continued this occupation up to the time of his admission to the Colony in the summer of 1912. His habits were moderate, he used tobacco in moderation and beer occasionally. There was no history of injury or of venereal infection.

History of the disease.—Epilepsy: his first seizure occurred at the age of 12 years. He was on his way from school, stopped to tie his shoe-string, and fell over in a convulsion. "Rush of blood to the head" was the assigned cause. Soon after this, myoclonic jerks and twitchings made their appearance. These gradually increased in severity and frequency and were especially

severe in the mornings. Myoclonia was usually greatly exaggerated before convulsive phenomena.

His general nervousness and the severe myoclonia were the reasons for his seeking admission to the Craig Colony in 1912.

Upon admission he was in good general health, and weighed 187 lb. He was "nervous and unstable." There were myoclonic jerks involving the head, trunk, upper and lower extremities.

His general intelligence was fair.

During the six years of his Colony residence, he has slightly deteriorated mentally, and has lost weight, but not rapidly. The myoclonia has greatly increased. The speech defect which was slight on admission has become intensified. There are exhaustion states following severe attacks of myoclonia, which at times may exist for days, and are quite general in their distribution.

It is definitely established that during the early years of his disorder he had severe grand-mal seizures, the tongue was bitten and there are scars about the face and scalp due to injuries received by falling during attacks. From 1912 to 1916 there are recorded occasional seizures, which appear on the regular Colony records, but most careful investigation fails to reveal, even from those in daily contact with him, that he has had attacks other than myoclonia in recent years. The present nurse in charge of his cottage, who has observed him for the greater part of five years, denies the occurrence of seizures, and states that the only manifestations of his illness are the "jerks and tremors." It is quite certain that the years of 1916, 1917 and 1918 have found him free from epileptic seizures, as he has been under close observation during that time. He has averaged during this period about two weeks per year in bed as a result of myoclonia, which made it impossible for him to get about, or even to feed himself.

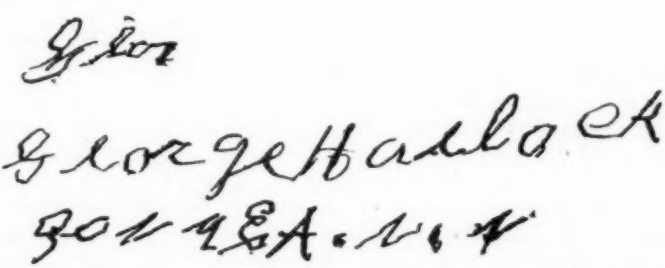
Physical examination. — Patient is a muscular, well-developed man. Station is steady and there is no Romberg symptom on closing the eyes. The spine is normal in contour and movements. The gait shows a slight clumsiness and awkwardness, especially on turning corners or facing about suddenly. At these times there is a slight tendency to uncertainty and the smooth harmony of balance and locomotion is somewhat disturbed. There is, however, no definite or distinct tendency to titubation and the slight disorder of gait and station might be regarded as merely clumsiness were it not for the existence of other and more characteristic cerebellar symptoms in the more delicate movements of the extremities.

The pupils are equal and react to light and accommodation. The ocular excursions are normal and there is no nystagmus (no history of diplopia). The other cranial nerves are negative. Fundi are normal.

Speech is of the cerebellar type, slow, of scanning character, and at times explosive. The tongue is protruded in the median line, without tremor. The innervation of the face, masseters and soft palate are normal. Frequently during the examination coarse myoclonic jerks and jumps occur in the extremities and trunk. These may be spontaneous but are particularly likely to

be initiated by any volitional effort or movement. At times the myoclonus occurs immediately after the cessation of a movement. Occasionally it is symmetrical on the two sides of the body, but both in the arms and legs independent myoclonic contractions are common. As in other cases the face is also occasionally involved.

There is no diminution of the gross motor power of the extremities and no evidences of muscular atrophy. On attempting to place the index finger on the tip of the nose a well-marked intention tremor is developed, which is more pronounced on the left side. This varies somewhat in intensity. At times there is present the coarse, slow, ataxic movement of dyssynergia which increases as the finger approaches the nose, on other occasions it has more the character of the typical intention tremor of multiple sclerosis. Evidences of dysmetria and typical adiadokokinesis are also present in both upper extremities, more particularly on the left side.



Patient's signature July 23, 1918, during "quiescent period" of the myoclonia.

In the lower extremities, evidences of cerebellar ataxia (dyssynergia and dysmetria) are also elicited when volitional movements are attempted, such as placing the heel upon the knee. There is no definite hypotonicity of the muscles of either arms or legs, and the Stewart-Holmes sign of hypotonia is absent.

Reflexes.—The arm jerks (supinator, biceps and triceps jerks) are present and active. The knee jerks and Achilles jerks are also present and active. There is no clonus. The skin reflexes are normal. The abdominal and cremasteric reflexes are elicited and the plantar reflexes are present and of the flexor type (no Babinski).

All sensations, both superficial and deep, are normal. The sense of position is quite normal in both upper and lower extremities. Abdominal palpation is negative and there is no enlargement of the hepatic areas, no abnormal pigmentary deposit of skin and sclera. The heart and lungs are normal. Urine is normal. Wassermann test of the blood is normal.

Comment.—In this patient, as in the others, there was difficulty in determining the approximate age of onset of the dyssynergia. The myoclonus-epilepsy is itself such a formidable condition and causes so severe a disturbance

of motility that it rather tends to mask the other disorder. With the exception of myoclonus-epilepsy and dyssynergia no other symptoms of organic disease of the central nervous system could be found. Symptoms indicative of multiple sclerosis, Friedreich's ataxia or gross cerebellar disease were not present. I would therefore regard the condition as one of dyssynergia cerebellaris associated with myoclonus-epilepsy.

There was no familial history of either disorder.

Case 4.

Summary.—A girl 15 years of age with nocturnal epilepsy, myoclonia and slight cerebellar dyssynergia of three years duration.

A young girl, 15 years of age, a private patient, first came under my care in July, 1921. The family history, so far as could be elicited, was negative. The father, a professional man, died as the result of an accident. Up to the time of his death, at the age of 45, he was in good health and had never presented any symptoms of nervous or mental disease. Mother is living and well. There is no history of epilepsy, myoclonia or other organic nervous disease in their forebears. The parents were not related.

Previous history.—Patient is an only child, born at term, normal pregnancy and non-instrumental delivery. She was breast fed for a year and with the exception of digestive disturbances in early life was a normal infant. No infantile convulsions. Menses began at the age of 12, and are quite irregular. She was sent to school at the usual age and is of average intelligence.

History of present illness dates back about three years and began with what have been called nightmares. She awakens in the night with a scream and passes into general convulsive attack with unconsciousness, frothing at the mouth, lasting several minutes, and followed by a period of mental confusion. At first these attacks were infrequent but later they increased in number and severity and now average once a week, sometimes oftener. Two years ago, that is, one year after the beginning of the nocturnal epileptic seizures, curious muscular jerks or twitchings made their appearance. At first these were not severe and were more marked on certain days, usually the day before the nocturnal seizures. The arms, legs and trunks were variously affected, later speech became involved and there was a slight tendency to myoclonic waves and twitching of the face. This tendency to myoclonus has gradually grown worse and on bad days it is rather difficult for her to walk alone and use her arms in dressing and eating—on these occasions she usually remains in bed.

On these bad days she sometimes falls to the ground while walking or standing, the legs suddenly giving way under her. On two occasions she had severe falls of this character, apparently associated with momentary blurring of consciousness. The patient insists, however, that in these falls there is no vertiginous sensation or momentary lapse of consciousness and she simply suddenly loses the ability to stand upright, her legs giving way under her. On one occasion when I was testing her gait and equilibrium she plunged quite

suddenly to the floor, all of a heap, striking her chin rather heavily. She immediately rose to her feet and was a little shaken from the sudden and severe fall, but as far as I could determine there was no obscuration of consciousness or pallor, as one observes in petit mal. The attack was caused by a sudden release of the muscular mechanism which underlies the posture of standing and is probably a massive expression of the myoclonia. In one of these she suffered quite a severe laceration of the scalp. As a rule the myoclonia is most severe a few hours before an attack and it is usually possible to foretell the epileptic seizures by this aggravation of the myoclonic phenomena.

A nurse who has slept in the room with the patient for a number of months gives the following description of the seizure. "An attack, or nightmare, is usually preceded by an unsteady afternoon and evening: a difficulty in eating and talking and an inability to walk unaided is shown at these times. The nightmare usually begins with a series of screams and guttural cries followed by a clenching of the teeth and tautening of all the muscles of the body. It is an impossibility to even move one of her fingers at this time. After an interval of ten minutes, or sometimes longer, there occur great convulsions of the body followed by profuse foaming at the mouth. If attempts to awaken the girl are successful she invariably seems dazed and very often lapses into childish manner of speech—sometimes half weeping—babbling some nonsense—repeating some word or sentence. Twice she has played with her toes and wept when I forced her to stop. Sometimes it takes more than half an hour to persuade her to lie down and sleep after a seizure. The day following she invariably has a dull nervous headache which lessens as the day goes on. The second day following is usually her strongest, brightest day. Nightmares occur on the average of once a week, but sometimes there are three, and rarely more than one in a night."

In addition to the nocturnal crises and the myoclonus, another group of symptoms has been observed, viz., a certain awkwardness or inco-ordination of movement. It is difficult to place the exact time of the appearance of these symptoms, for the reason that the night crises and myoclonia have dominated the clinical picture. It is quite certain however that they have been in existence for the two years, gradually growing more severe. (The mother states that even before the appearance of the nocturnal seizures the movements of the extremities were somewhat clumsy and awkward, and this was especially noticeable in her table manners. This clumsiness was not taken seriously at the time and was regarded simply as a peculiarity of childhood, which would pass off in the course of time.) There is a certain awkwardness in walking and a tendency to balancing on rising from a sitting posture. There is an awkwardness and uncertainty in the use of the arms and hands and a thickness and uncertainty of articulation.

Mentally she is bright and intelligent, but memory is not so good as formerly, and she is at times peevish and irritable, especially on the bad days

when the myoclonia is exaggerated. She is subject to headaches after the nocturnal crises, but not at other times, and is in good general health.

Physical examination.—She is well developed, of average height and muscular development. Seated in a chair and while conversing certain of the muscles of the face, trunk or arms will exhibit a quick muscular contraction, producing a marked locomotor effect. During the interval between these moments of myoclonia her expression and posture are perfectly normal. The myoclonic jerks are frequently initiated by a voluntary movement, such as reaching for some object, an effort to speak or a movement of the head. Usually there is a single jump or jerk and then rest. Occasionally a series of such movements occur and the trunk or limbs are thrown about in a rather bizarre manner.

The pupils are equal and react to light and accommodation. There is no nystagmus. The optic nerves are normal. All of the motor cranial nerves are normal. The articulation is slow, jerky and uncertain. In addition to occasional myoclonic speech disturbance there is slight dysarthria of cerebellar quality.

The upper extremities are normal in their gross motor power and there is no tremor of the outstretched hands. There is however a slight ataxia on the finger-nose test, and a distinct tendency to dysmetria and a very typical dysdiadokokinesis of both arms and fingers. There is also slight hypotonia of the musculature. The Stewart-Holmes sign of hypotonia is absent.

On arising from a sitting posture there is a certain static ataxia and a tendency to balance the trunk. In walking the legs are spread somewhat and there is a moderate degree of cerebellar disorder of progression which is especially obvious on turning corners or turning round suddenly.

In the recumbent posture the heel-knee test shows a moderate degree of dysmetria. There is however no obvious hypotonia. On elevating both arms and legs in the recumbent posture, with the eyes closed, there is no ataxia.

The tendon reflexes of both upper and lower extremities are present and equal on the two sides, viz., the supinator, biceps, triceps, knee jerks and Achilles jerks. Sensation, both superficial and deep, is normal. The spinal column is of normal contour and there is no deformity of the feet. The heart and lungs are normal. Abdomen is normal on palpation and percussion. There is no enlargement of the liver. There is no pigmentation of the skin or cornea. Urine shows no abnormality.

Comment.—The case is of interest as representing a somewhat earlier stage of the dyssynergia cerebellaris myoclonica. Here again there is no family history of myoclonus-epilepsy or of a cerebellar disorder.

Remarks.—The association of cerebellar dyssynergia and myoclonus-epilepsy in this group of cases suggests the occurrence of two independent nervous disorders in one individual. Combined forms of the various congenital, familial and hereditary types of nervous disease are

by no means uncommon, and these associations are especially frequent in the spinal and cerebellar groups of system diseases. For example, Jendrassik [14], who has devoted particular attention to this subject, records many such combinations. He mentions a combined form of Friedreich's ataxia and muscular dystrophy, of spino-cerebellar ataxia and muscular dystrophy, and of Friedreich's ataxia with Huntington's chorea. He also cites the occurrence of myoclonus in association with optic atrophy. André-Thomas [3] has also described a combined form of olivo-ponto-cerebellar atrophy and spinal ataxia.

Such combinations, however, are rare, and show merely a predisposition to the two disorders in the same individual, and would not necessarily indicate any essential relationship between them.

On the other hand, little is known at the present time of the pathology and localization of myoclonus. Its occurrence, therefore, in conjunction with a special type of cerebellar disease, is not without interest, and may have a deeper pathological significance than would appear.

Sioli [19], in a histological study of one of Rechtenwald's cases of myoclonus-epilepsy, found rather widely disseminated histological changes.

In the cerebellum there were enormous collections of lipoid material in the region of the corpus dentatum in the white matter of the cerebellar hemispheres and in the pons. There was also a considerable increase in the number of glia cells. The cells of the corpus dentatum were normal. In the cerebral cortex there was also an excessive deposit of lipoid material in the ganglion cells and glia. The glia cells and fibres were increased in the marginal layer. The cortical changes were all more conspicuous in the region of the central convolutions. The only alteration noted in the spinal cord was atrophy of Hellweg's triangular bundle (olivo-spinal tract).

In the present state of our knowledge, while these two disorders may be regarded as independent manifestations of two distinct disorders in the same individual, the possibility of a closer pathological association should be borne in mind, and the cerebellar mechanism should be carefully considered, both clinically and pathologically, in all cases of so-called myoclonus-epilepsy.

Chapter III.—Dyssynergia Cerebellaris Myoclonica, associated with Friedreich's Ataxia.

Clinical Report of Two Cases, One with Histological Study of the Central Nervous System.

In the previous chapter there was described a group of cases, characterized by progressive dyssynergy, associated with myoclonus-epilepsy. In none of these cases was there any history of an hereditary or familial tendency to either disorder.

In the present chapter I shall present another group of cases in which myoclonus-epilepsy and dyssynergia cerebellaris progressiva were associated with symptoms of Friedreich's ataxia. It is well known that the various cerebellar atrophies and degenerations are not infrequently associated with evidence of spinal cord involvement. Friedreich's hereditary spinal ataxia and Marie's so-called hereditary cerebellar ataxia are classical examples of this association, and may exist alone or in combination. The progressive cerebellar dyssynergia is apparently no exception to this rule—it may exist alone or in combination with spinal cord involvement of the Friedreich's type.

Case 5.

Summary.—A man, aged 36, with symptoms of myoclonus-epilepsy since his twenty-first year. Previous to this there were symptoms of Friedreich's ataxia and progressive cerebellar tremor, which steadily progressed. Pathological study shows the typical spinal lesions of Friedreich's ataxia associated with a primary atrophy of the efferent dentate system of the cerebellum, viz.: considerable diminution in number and extensive atrophy of the cells of the dentate nucleus, with secondary atrophic changes in the superior cerebellar peduncles.

The patient, a man, aged 36, single, was admitted to my service at the Montefiore Home and Hospital on April 15, 1918.

Family history.—His father is living and well. Mother died at 51 from heart disease. The parents were not related. One brother died at age of 2 from pneumonia. He has two other brothers and one sister living and in good health. There is no history of tuberculosis, diabetes, epilepsy, or other nervous disease in the family, with the exception of a twin brother who is suffering from the same malady, and whose case will be presently described.

Previous history.—Had measles in childhood. Was always moderate in the use of alcohol, coffee, and tea. Smoked cigarettes in moderation. Denies venereal infection. No history of trauma.

Present illness.—Began fifteen years ago with attacks of unconsciousness

and convulsive seizures. Patient believes these attacks were caused by fright following a burn. At the time of the accident he was incapacitated for some weeks. The epileptic seizures increased in frequency, and he sometimes fainted on the street and was carried home. A few months later there appeared coarse jerky movements of the extremities. The myoclonus jerks or starts came on gradually and steadily increased in severity and extent.

Even in the earlier juvenile period it was noticed that his manner of walking was awkward and uncertain, and this steadily increased. For the past ten years walking has been unsteady and difficult, speech has been slow and there has also been some uncertainty in the use of the hands. He thinks that part of his inability to walk is due to the fear of falling on account of the myoclonic jerks and starts, which develop on making the slightest effort.

For the past seven years his speech has been quite indistinct and for about the same period of time he has had a marked volitional tremor of the hands on attempting to grasp an object. For three or four years he has suffered from very extreme weakness and unsteadiness of lower extremities and is unable to walk.

During the whole course of the disease there have been no subjective sensory disorders, such as pains or paræsthesias, and there has been no difficulty with the control of the sphincters of the rectum or bladder. States that he has not suffered from headache. He now wears dark glasses constantly, because any bright light increases the tendency to myoclonia. Hearing is not impaired. There are slight evidences of mental impairment and deterioration. His memory is poor, and his mental processes slow. He is querulous, irritable and easily excited.

Physical examination.—Patient is confined to a wheel chair, and is unable to stand or walk without assistance. He wears dark glasses and always seeks a quiet corner of the ward where there is no noise and no excitement. The slightest muscular effort brings on a series of severe myoclonic jerks or spasms which throw him brusquely to and fro in his chair or bed. Talking has the some effect as have also noises or any excitement. As a result of these continuous motor shocks the patient shuns conversation and any social contact. He wishes to be left alone and in quiet.

His speech is slow and typically scanning, and at times uneven and explosive. It is often interrupted by myoclonic jerks and starts in the trunk and extremities, causing sudden shock-like movement. His head is frequently thrown backwards in sharp retro-colic clonus, associated with quick flexor movements of extremities. This larger myoclonic phenomenon is frequently repeated during the course of the examination. Occasional facial grimaces of myoclonic type are also noted. These movements are all accentuated by speech and voluntary movement. The clonic movements are said to occasionally persist in sleep, although in much diminished form. There is a typical and very marked intention tremor of both upper extremities which is associated with dysmetria and dysdiadokokinesis. On attempting to carry the finger tip to the nose the hand is thrown into a violent ataxic tremor similar to

that observed in an advanced case of multiple sclerosis. All tremor movement of the arm immediately ceases in a relaxed or recumbent posture. There are, however, occasional myoclonic starts at rest, which are increased by some one entering the examination-room suddenly. The musculature of the arms is hypotonic and the Stewart-Holmes sign of hypotonia is present in both upper extremities.

Asthenia is present in both arms, viz., when the attempt is made to extend the arm from the body in abduction and maintain the abductive extremity in this position. It can be maintained for a very brief period only, the arm falling to the side, a symptom which may be observed in the intention tremor of multiple sclerosis (asthenia or astasia).

The arm jerks are difficult to elicit and the supinator, biceps and triceps jerks are not definitely present. The abdominal reflexes are not elicitable. The cremasteric reflexes are present on both sides. The plantar reflexes are present but diminished and of the flexor type (no Babinski).

The movements of the lower extremities are extremely ataxic, but in addition to this there is on performing the heel-knee test a very marked intention tremor of both lower extremities. The legs are markedly hypotonic. The knee jerks are absent and the Achilles jerks cannot be elicited. He can stand and walk only with assistance. The Romberg symptom, owing to the extreme ataxia, cannot be tested. In walking, the legs are lifted high and thrown outward in a very ataxic and uncertain manner and resemble the gait of advanced locomotor ataxia. There is no definite spinal curvature but a tendency to pes cavus is present. There is general emaciation and wasting of the musculature but no localized degenerative atrophies.

Sensation.—There is a marked disturbance of the deep sensibility of the lower extremities, and loss of the sense of position. The deep sensibility of the hands is also affected and there is a loss of the sense of position in the hands and fingers. The superficial sensibility to touch, pain and temperature is diminished over the more distal portion of the lower extremities, but is nowhere abolished. The superficial sensation of the upper extremities is fairly well preserved: the only change being diminished acuity of perception over their distal portion. Myotatic irritability of the muscles is active and somewhat increased.

The pupils are equal and react to light and accommodation. Ocular excursions are normal. There is no nystagmus. Hearing is normal. The tongue is protruded in the median line; no atrophy and no tremor. Innervation of the palate is normal. The pharyngeal reflex is active. Facial innervation is normal as well as that of the muscles of mastication. Jaw jerk is not elicitable. The ophthalmological examination by Dr. Tenner revealed no abnormalities. The fundi were negative.

The heart and lungs are negative. Systolic blood pressure 110, diastolic 70. Wassermann test of blood negative. There are no pigmentary deposits of the skin or sclero-corneal margin.

Since his admission to the hospital patient has had very active myoclonia,

The head is thrown suddenly backward, the face is convulsed with occasional myoclonic waves, the arms flex and jerk, the trunk is thrown forward and the legs are flexed and tossed about in a wild and random manner. There are one or two discharges and then a period of rest, although these rest intervals are of very short duration. It is necessary to bind patient with a sheet to the chair, otherwise he would be thrown out, and even with this precaution the wheel-chair rolls around in a wild and reckless manner. This condition is very much aggravated by removal of the dark glasses and the excitement of any examination. Any visit or conversation induces an extreme degree of myoclonia. This condition gradually grew worse, the patient dying of exhaustion a few weeks later.

Comments.—This case differs from those described in the previous chapter in two respects, viz.: the association with Friedreich's ataxia and the familial incidence, a twin brother suffering from the same disease. The first symptoms of the cerebello-spinal disorder made their appearance in the early juvenile period. The myoclonus-epilepsy did not become manifest until the twenty-first year. The presence of the spinal symptoms of Friedreich's ataxia would mask many of the typical symptoms of cerebellar disease. The existence, however, of marked intention tremors and the scanning speech show very clearly the existence of cerebellar involvement (cerebellar tremor) and this was amply confirmed by histological examination.

The central nervous system was subjected to careful histological examination, and revealed the typical spinal lesion of Friedreich's ataxia, together with primary atrophy of the efferent dentate system of the cerebellum, viz.: diminution in number and atrophy of the dentate nucleus with atrophy of the superior cerebellar peduncles. (See report of histological examination.)

Case 6.

Summary.—A man, aged 29 years, the twin-brother of Case 5. Symptoms of dyssynergia cerebellaris and Friedreich's ataxia for the past ten years, gradually increasing in severity. Myoclonia made its appearance five years ago and epilepsy is only of three years duration.

History.—Patient is 29 years of age, the twin-brother of the case just described. He assists his father in business, who is a tailor by occupation. He resembles very closely his twin-brother in face and stature, and states that he has had symptoms of the same disorder for the past ten years or more. Before that he could run and do anything that other boys were able to do. The condition has been very slowly progressive.

It is characterized by thickness of speech, ataxia and intention tremor of the hands and arms, a certain awkwardness of the movements of walking and running and the symptoms of myoclonus epilepsy. At times there is a lateral tremor movement of the head.

For the past five years he has been subject to nervous shocks or starts, which at first occurred only three or four times a day, but have steadily increased in frequency and intensity. These attacks of myoclonus may

occur spontaneously but more frequently accompany attempts at voluntary movement. They have never approximated in severity those occurring in his brother's case. Efforts to talk are frequently accompanied by clonic "starts" and "jumps" (myoclonus), usually only a single movement which may involve the arms, face, neck, trunk and sometimes the legs.

Three years ago he had his first epileptic seizure. At first these attacks were infrequent but have gradually increased in frequency and during the past year have occurred as often as twice a week. In the seizures he is momentarily unconscious and falls to the ground. The obscuration of consciousness is, however, of very short duration. It is as if his legs give way under him, and a darkness comes over his eyes, but he gets up immediately and goes on with what he was doing. There is no biting of the tongue, vomiting, or incontinence of the sphincters, and so far as could be learned no convulsive manifestations. This is a type of seizure which I have referred to the static or posture system of motility—static epilepsy. It is characterized by a sudden loss of control of the static or posture mechanism of standing with transitory obscuration of consciousness and without the usual convulsive manifestations of epilepsy. During the past few years his memory is not so good. There is no history of headache or diplopia, and the special senses are unimpaired. There is no vertigo; no tinnitus aurium; no attacks of nausea or vomiting.

Physical examination (April 28, 1918).—He is of medium height, slenderly built, and bears a very strong resemblance to his twin-brother. The pupils are equal and respond promptly to light and accommodation. The ocular excursions are normal and there is no nystagmus. Motor innervations of the face, tongue, soft palate and muscles of mastication are normal. The fundi are negative. At times, in certain positions a lateral tremor of the head develops.

The speech is dysarthric; rather slow and scanning and at times of a slightly explosive quality. The tongue is protruded in the median line; there is no atrophy or tremor and the palatal innervation is normal. The jaw jerk is not elicitable. Efforts to speak are often accompanied by mild clonic starts or jumps, usually, only a single one, which may involve the face, neck, arms, trunk and sometimes the legs. The station shows a slight unsteadiness but there is no special increase in this slight disorder of equilibrium on closure of the eyes. There is slight awkwardness in walking, especially on coming to a sudden stop or on turning corners. There is a moderate scoliosis in the mid-dorsal region, with a slight convexity towards the left. There is no muscular atrophy of the trunk, arms or legs and no fibrillary twitching. There is no deformity of the feet (no pes cavus). The gross motor power of the arms is normal. The movements, however, are awkward and ataxic and on attempting to carry out a volitional movement a typical intention tremor develops, which is more marked on the left side. The musculature of the upper extremities is moderately hypotonic and the Stewart-Holmes sign of hypotonia is present. (Handwriting, p. 509.)

The arm jerks cannot be elicited (supinator, biceps or triceps jerks). There is typical adiadokokinesis of both upper extremities. There is no loss of

gross motor power in the lower extremities and no evidence of muscular atrophy. There is a moderate degree of hypotonicity of the musculature and the knee jerks and Achilles jerks are absent. The plantar reflex is of the flexion type (no Babinski). The other skin reflexes, abdominal and cremasteric, are present and equal on the two sides.

On attempting to carry out the heel-knee test there are distinct evidences of dysmetria and of an ataxic disturbance, suggestive of the intention tremor but not so distinct as in the upper extremity.

Sensation appears to be normal. The superficial sensibility to touch, pain and temperature is intact, and the deep sensibility of the toes is well preserved. The muscle pain sense on pressure is well preserved.

Comment.—This case, like that of his brother, presents symptoms of dyssynergia cerebellaris progressiva, myoclonus-epilepsy and Friedreich's ataxia in combination.

According to the history, symptoms of cerebellar tremor and Friedreich's ataxia made their appearance some years after the development of myoclonus or epileptiform seizures. It is interesting to note that the epileptiform seizures correspond to what I have termed the static type of epilepsy, viz., a sudden loss of postural or static control without convulsive manifestations.

That the disorder in which we are interested has also been encountered by other observers is shown by the following extract from a case reported by Boschi [5] as ataxie héréditaire; paramyoclonus multiplex, type Unverricht.

The patient was a man 46 years old, who at the age of 15 was subject to convulsive seizures and myoclonus, associated with a well marked ataxic disorder, which had gradually shown a tendency to progression. There was in addition a very marked familial predisposition to the disease. The grandfather, father, and four brothers were all sufferers from the same malady.

Physical examination shows an ataxia of gait, of the movements of the trunk and of the arms. In both upper extremities there is present an intention

tremor. The speech is slow and dysarthric (bradylalia). The knee jerks are exaggerated and there is clonus. The pupillary reflexes are present but slow. The optic nerves are normal. There is a moderate mental defect and typical myoclonus of the Unverricht type.

The case, which is very briefly recorded, would seem to resemble Cases 5 and 6 of my series: dyssynergia cerebellaris myoclonica with Friedreich's ataxia. It is interesting because of the striking hereditary nature of the malady, which was absent in my series of cases.

Undoubtedly other cases have been observed and recorded, but, with this single exception, I have encountered in the literature no other examples in which myoclonus-epilepsy was observed in conjunction with evidence of a cerebello-spinal disorder.

Pathological Study of Case 5.

Autopsy Report.

The autopsy was performed thirty-eight hours after death, by Dr. J. H. Globus, Pathologist to the Montefiore Home and Hospital. His report is as follows:—

The body was that of a poorly developed male, 160 cm. in length. Skin thin, dry, pale, with several decubitus ulcers over lower sacrum, under the anterior superior spine of ilium and over internal tuberosity of right tibia.

The regional lymph nodes were normal in size and slightly palpable.

External genitalia were undeveloped, otherwise normal.

Scalp was free of hair and mottled with numerous small superficial ulcerations. Rigor mortis had disappeared.

On incising abdominal wall, a very small quantity of subcutaneous fat was found. On opening peritoneal cavity no adhesions; no fluid encountered.

Abdominal viscera were found in normal relation to each other and in normal position. No fluid and no adhesions in thoracic cavity.

Lungs, rather voluminous and collapsed only to slight degree. The pericardial cavity was opened after the removal of a thin layer of fatty thymus, and was found free of fluid and adhesions.

Heart, small sized and contracted; weighs 190 gm.

Length of heart, 9 cm.

Muscle bundles of fair size and consistency and of greyish-brown colour. Sub-epicardial fat decreased in amount. All valves intact and normal in size and thickness.

Lungs: Right weighs 600 gm., voluminous, covered by smooth

glistening moist pleura of dark purple-greyish colour. It is interrupted here and there by accumulation of exogenous pigment. On cut section presents a dark purple surface throughout the three lobes of the lung, markedly increasing in consistency, and with numerous dark brownish patches scattered throughout the surface. On pressure, a frothy pinkish fluid is expressed from the alveoli bronchi. No scars, no calcareous deposits are made out.

Left lung: Weighs 500 gm.; presents a similar picture throughout its lower lobe, which is also markedly congested, showing several small patches of consolidation and permits the escape of a pinkish, sero-sanguineous fluid from the alveoli. The upper lobe of left lung is well aerated, of a bright pink colour, collapsed, showing no pathological changes whatsoever.

Liver: Weighs 1,100 gm.; below average in size; surface covered by thin, glistening capsule of a greyish-yellow colour. Cut surface is of a greyish-cream colour. Consistency markedly reduced, lobulation indistinct. Iodine test negative.

Spleen: Weighs 100 gm., small size capsule, thickened and wrinkled. On cut section surface is of a brownish-red colour. Trabecular and Malpighian corpuscles are distinct.

Kidneys: right weighs 120 gm.; left weighs 110 gm.

Both present similar picture. They are of small size. Capsule strips with ease, leaving a smooth greyish-yellow surface. Consistency markedly diminished. On cut section medulla is of normal size, cortex reduced in amount. Aside from a pale yellow colour, no definite pathological changes.

Adrenals: right rather well preserved; left shows a soft medulla with a congested central vein.

Gastro-intestinal tract: the stomach is normal.

Small intestines are collapsed; mucosa shows several patches of marked congestion, otherwise normal.

Large intestine is filled with greenish fluid, on the removal of which a thickened mucosa with numerous small, deep, clean ulcers scattered throughout the length of the large intestine. The ulcers measure from 3 to 5 mm. in diameter, irregular in outline, rather deep seated, base clean and glistening. They become more numerous in the lower portion of the large intestine.

Pancreas is normal.

Blood-vessels are rather strikingly free of arteriosclerotic changes, very elastic but somewhat reduced in circumference.

Bladder is markedly distended, muscular walls stretched, mucous membrane pale, showing a few petechial hæmorrhages.

Testes are small and soft; tubules easily pulled out of supporting tissue.

Prostate is apparently normal.

Neck organs: Right thyroid is twice the size of left, consistency normal. Three parathyroids measuring 5 mm. in diameter, oval in outline, were removed.

Larynx, normal.

Œsophagus, normal.

Brain is of moderate size, shows no apparent reduction in the prominence of the gyri. Several white opacities are seen in the meninges of the cerebral hemispheres. The vessels of the cortex and circle of Willis are normal.

Spinal cord was found to be of average size. Meninges appeared normal; there was no section evidence of degenerative atrophy in the posterior columns, but no other changes could be made out.

Microscopical examination of the viscera (Dr. J. H. Globus):—

Lungs: Alveoli are crowded with red blood cells and polymorphonuclear leucocytes. The lining epithelium is desquamated, the blood-vessels in the walls are markedly congested. Bronchial tubes have lost their membrane lining, the lumen filled with desquamated epithelium—polymorphonuclear leucocytes. The wall shows marked congestion of blood-vessels.

The liver is normal.

Spleen: trabeculæ prominent, walls of blood-vessels markedly thickened, Malpighian corpuscles in large numbers, fine size, blood-vessels engorged.

Kidneys, normal.

Pancreas, normal.

Adrenals, normal.

Thyroid: alveoli filled with pink staining colloidal matter. Lining epithelium of alveoli extremely thin. Their cellular structure can hardly be made out.

Spermatic cord, normal.

Prostate: alveoli are few in number, few concretions present.

Colon: several small abscesses opening on the surface of the mucosa are found extending deep into the submucosa.

Skeletal muscles: muscle fibres small in calibre, fragmented striations indistinct, nuclei few in number and pyknotic.

Anatomical Diagnosis.

Hypostatic congestion of lungs, lobular pneumonia, pulmonary œdema, fibrosis of spleen and ulcerative colitis.

Histological Examination of the Central Nervous System.

Blocks of tissue were taken from the cerebral cortex, the cerebellar cortex, the dentate nucleus, and the spinal cord for alcohol fixation and the Nissl method.

The brain and cord were fixed in formalin and various levels removed for cellular stains (toluidin blue, hæmatoxylin eosin). The remainder was then placed in Müller's fluid in preparation for the Weigert-Pal method.

Microscopic sections were prepared in the laboratory of the Montefiore Home and Hospital. The larger Weigert-Pal sections were made in the neurological laboratory of Columbia University, and I am much indebted to Professor Tilney for extending to me the courtesy of the laboratory in the preparation of this work.

Spinal cord: The large multipolar cells of the anterior horns are normal, at the cervical, dorsal and lumbo-sacral levels. The anterior roots are normal. The cells of the columns of Clarke in the dorsal region show signs of atrophy and are diminished in number.

The posterior columns show advanced atrophic change which may be traced from the lower lumbo-sacral levels to the nuclei of Goll and Burdach in the lower portion of the medulla (fig. 1).

The columns of Goll are almost completely atrophic, while those of Burdach show a lesser degree of involvement. The posterior cornua appears somewhat reduced in size. The fibres of Lissauer's tract appear fairly well preserved.

There are marked evidences of atrophy in the lateral columns, corresponding to the ventral and dorsal-cerebellar tracts (direct cerebellar and Gowers' tract). There is a lesser degree of diffuse atrophic change in that portion of the lateral columns corresponding to the crossed pyramidal tracts and the spino-thalamic tracts. In the area occupied by the crossed pyramidal tracts this is not clearly defined and as the Babinski reflex was absent it would seem likely, as suggested by Marie, that these atrophic fibres belong to other non-pyramidal systems. This would seem all the more probable as these slighter evidences of atrophy are limited to the cervical and dorsal regions and are scarcely at all demonstrable at the lumbar levels.

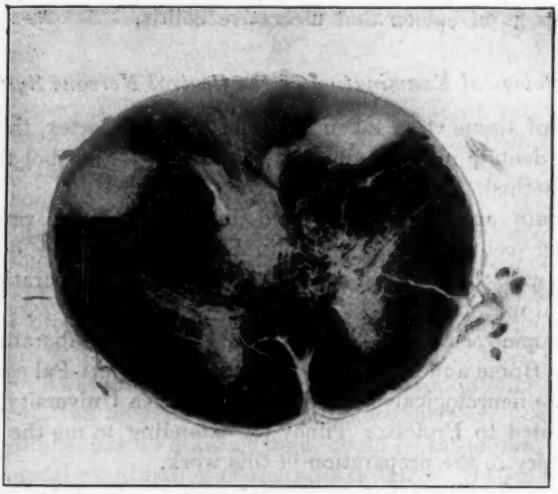


FIG. 1.—Cross section of the medulla oblongata, at the level of the pyramidal decussation. Weigert-Pal stain, showing atrophic changes in the tracts of Goll, Burdach and the spino-cerebellar tracts.

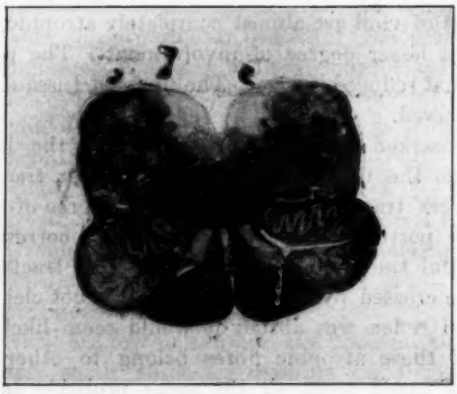


FIG. 2.—Medulla at the level of the inferior olive, showing atrophy of the spino-cerebellar tracts. The pyramidal tracts, the fillet, and the inferior olive and olivo-cerebellar fibres are well preserved.

In the anterior columns there is some marginal pallor, especially in the region of Helweg's tract. The direct pyramidal tracts are not atrophic.

The chief evidences of system involvement are therefore confined to the columns of Goll, Burdach, and the ventral and dorsal spino-cerebellar tracts. In both the cervical and dorsal regions there are, however, slight but rather diffuse atrophic changes.

There are no vascular lesions other than slight secondary thickening in the sclerosed areas, and no evidences of inflammatory lesions. The meninges are slightly thickened in the region of the sclerosed tracts but show no acute changes. The posterior roots show evidences of atrophy. The anterior roots are well preserved.

There is a combined degenerative atrophy of the postero-lateral columns of the cord. Fairly complete of the column of Goll, partial of the column of Burdach; and also of the direct cerebellar tract, Gowers' tract and of the columns of Clarke (cells and fibres). In addition to this there were slight diffuse atrophic changes in the lateral columns. The pyramidal tracts were not atrophic, which is supported by the clinical findings (absence of the Babinski reflex).

Medulla oblongata (fig. 2).—The cranial nerve nuclei of this region (12, 10, 9 and 8) are normal. The pyramids are of normal size and configuration, and there is no evidence of pallor in the Weigert-Pal specimens. The fillet is also normal in size and appearance. The direct cerebellar tracts show atrophic changes as they pass into the corpora restiforme.

The nuclei of Goll and Burdach as well as the fibræ arcuatæ internæ are well preserved.

The inferior olives and accessory olives are normal in size, general appearance and cellular content. The olivo-cerebellar fibres are well preserved.

The descending roots of the trigeminus and the solitary fasciculus are not atrophic.

The fasciculus longitudinalis posticus is normal.

The only noteworthy pathological alterations in this region are the evidences of atrophy posteriorly in the lower levels of the medulla where the tracts of Goll and Burdach pass into their respective nuclei and laterally correspond to the direct cerebellar tracts.

The large sensory and motor projection systems and the inferior olivary complex and its fibres of communication with the cerebellum are normal.

FIG. 3.—Vertical section through cerebellum and pons, at the level of the intrinsic cerebellar nuclei. The peduncles and arbor vitæ of the cerebellum are well preserved.

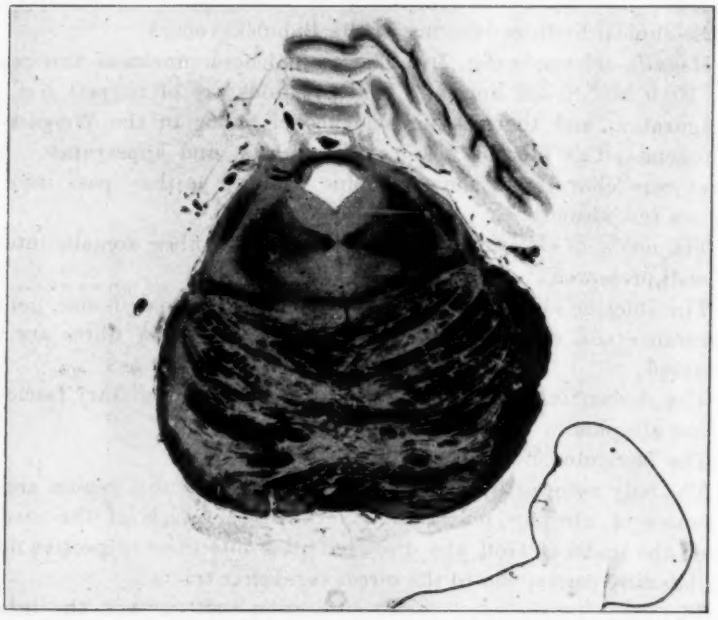


FIG. 4.—Cross section through the pons at the level of the trochlear decussation. Note the marked reduction in the fibres of the superior cerebellar peduncles and especially the atrophic changes in the region of the peduncular decussation. The pontine fibres constituting the stratum superficiale and the stratum profundum are well preserved.

Pons varoli (figs. 3 and 4).—The pontine nuclei, both ventral and dorsal groups, are normal and show no reduction in size or number of the cells. The fibres of the stratum superficiale pontis and the stratum profundum pontis and their continuation in the middle cerebellar peduncles are free from atrophic changes.

The corpora trapezoidii and the superior olives are normal. In higher levels of this region the superior cerebellar peduncles show distinct evidences of atrophy and considerable reduction in size. There are also atrophy and evidences of a marked diminution in the number of fibres at the decussation of the superior cerebellar peduncles.

The cranial nerve nuclei of this region are normal (eighth, seventh, sixth and fifth nerves).

The lateral fillet is normal and the fasciculus longitudinalis posticus is well developed on both sides. The essential pathological change in the pons is an atrophy of the superior cerebellar peduncles. The other structures of the pons, including its peduncles and the pontine nuclei, are normal in size and appearance.

Mid-brain (figs. 5 and 6).—This region reveals the atrophy of the fibres of the superior cerebellar peduncles by the diminished decussation of the peduncles which is so characteristic a feature of these levels. The lateral lemniscus and both anterior and posterior colliculi are normal in appearance.

In the higher levels the ventral decussation of Forel appears apparently in full strength; so while there is a distinct and well-marked atrophy of the fibres passing between the dentate nucleus and the red nucleus this is apparently not continued over into the rubro-spinal tract, which receives further confirmation by the preservation of the cells of the red nucleus.

The various oculo-motor nuclei and root fibres are normal. The cells of the locus niger and of the nucleus ruber appear normal and so far as could be determined there was no reduction in their size or number. Especially interesting is the normal condition of the long projection tracts of the crus cerebri, viz., the fronto-ponto-cerebellar, the pyramidal and parieto-temporo-occipito-cerebellar tracts. The ponto-cerebellar tracts show no evidences whatever of atrophy or pallor.

The only noteworthy change in the mid-brain was the well-marked atrophy of the fibres which enter into the decussation of the superior cerebellar peduncles. I would also emphasize the normal appearance of the connections between the cerebrum and the cerebellum, viz., the fronto-ponto and the parieto-temporo-occipito-ponto cerebellar tracts.

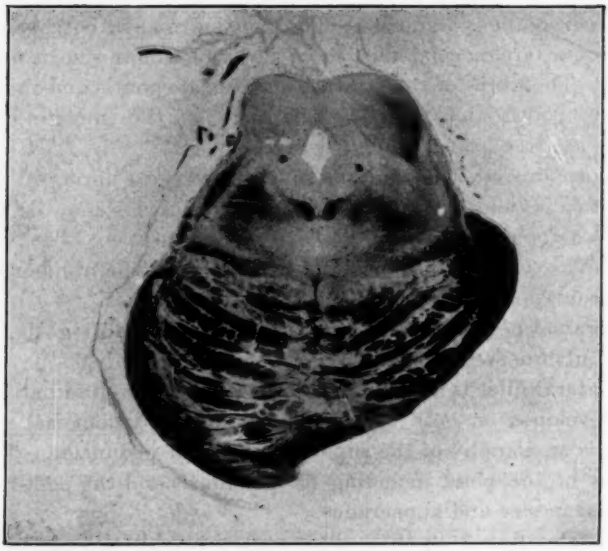


FIG. 5.—Cross section through the inferior corpora quadrigemina, at the level of transition from pons to midbrain. Note the marked reduction in size and the atrophic changes in the region of the superior cerebellar peduncles and their decussation. The stratum superficiale and stratum profundum of the pons are well preserved.

FIG. 6.—Cross section through the superior corpora quadrigemina. The long projection fibres of the crura cerebri are well preserved, including the fronto-cerebellar and the parieto-temporo-occipito-cerebellar tracts. Note diminution of peduncular fibres in the region of the red nucleus.

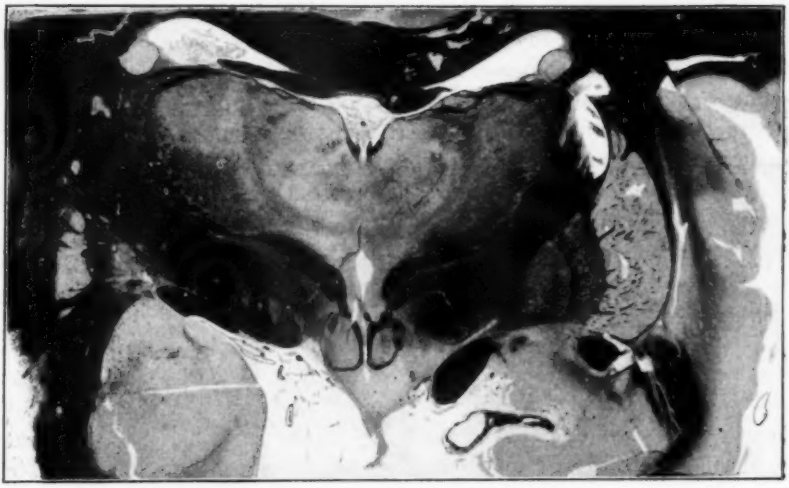


FIG. 7.—Cross section through the interbrain showing the corpus striatum and optic thalamus.

FIG. 8.—Section through the cerebellar hemisphere showing normal arbor vitæ and cerebellar cortex.

Interbrain (fig. 7).—Sections of the interbrain reveal no abnormalities of the optic thalamus or of the corpus striatum. The cells of the locus niger and nucleus ruber are well preserved and are normal in number and appearance as are those of the corpus subthalamicum. There is no apparent atrophy of the fibre systems of either the optic thalamus or corpus striatum.

Cerebral cortex.—Sections of the cerebral cortex from the frontal, parietal, temporal and occipital areas were examined by cellular and fibre stains and revealed no noteworthy abnormality.

The meninges and blood-vessels showed no special pathological changes.

Cerebellum (fig. 8).—The general histological appearance and markings of the cerebellum are quite normal. The arbor vitæ is well developed and the grey matter of the cortex is normal. The molecular, Purkinje, and granular layers of the cerebellar cortex present a normal histological appearance. The large cells of Purkinje are normal in size and number.

Vertical sections were made through the cerebellum including the structures of the pons and medulla oblongata. In these sections were included the intrinsic nuclei of the cerebellum.

The convolutions of the nucleus dentatus appear a little less voluminous than is normal and the cerebellar olive gives the impression of being somewhat reduced in size. The nucleus emboliformis, globosus and tecti are present and of normal size and appearance.

Histological study shows a considerable atrophy and diminution in number of the cells of the dentate nucleus (figs. 9, 10, 11, 12). Many of these cells appear shrunken, rounded and atrophic, and a large number have completely disappeared. An attempt was made to estimate in some degree the cellular loss by making comparative cell counts with normal specimens. By this method the loss in the number of cells varied from one-half to one-third.

Because of the small size of the other intrinsic nuclei of the cerebellum (nucleus tecti, emboliformis and globosus) it was not possible to make the same accurate estimation of the cell content as in the case of the larger nucleus dentatus.

A study of the sections, however, revealed little, if any, evidence of cellular loss or atrophy. The cells of the nucleus tecti especially were numerous and well preserved. The chief cellular changes were confined to the convolutions of the cerebellar olive (corpus dentatum). The meninges and vascular structures of the cerebellum were normal. Only

within the nucleus dentatum did the blood-vessels show some secondary thickening.

The cerebellum was, therefore, normal in its histological appearance, with the sole exception of an extensive and well-marked atrophy of the cells of the dentate nucleus.

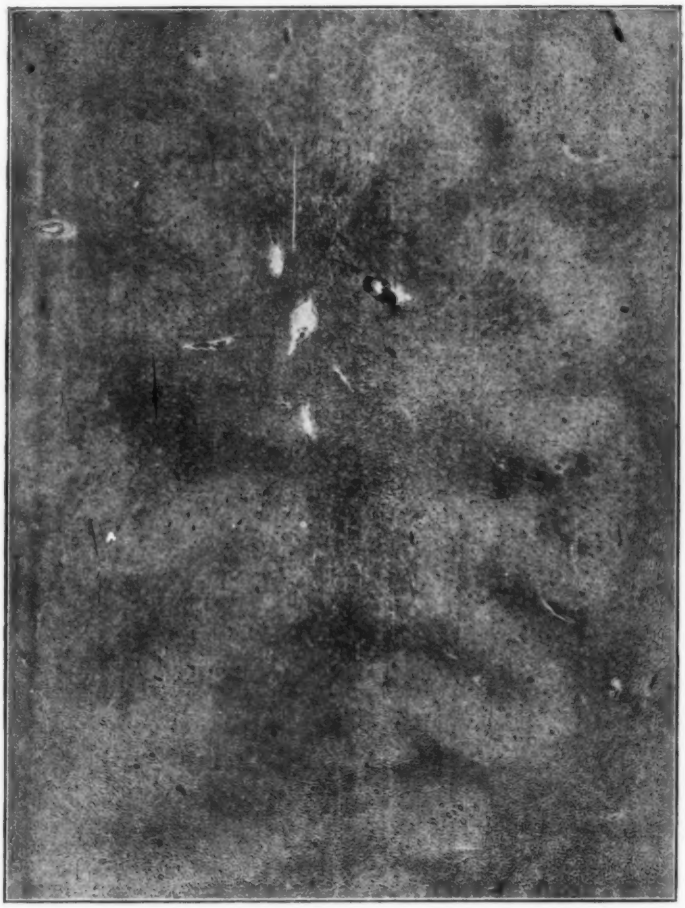


FIG. 9.—Dyssynergia cerebellaris myoclonica. Primary atrophy of the dentate system. Hæmatoxylin-eosin stain, section of the corpus dentatum, showing the convolution of the cerebellar olive and the great reduction in size and number of the cells of the corpus dentatum.

Summary of Pathological Changes.

The histological study of the central nervous system in this case showed the typical spinal cord changes of Friedreich's ataxia. There was atrophy of the posterior columns of the cord which could be traced

FIG. 10.—Normal control: same magnification and stain as fig. 9. Section through the corpus dentatum, showing convolution of the cerebellar olive and normal appearance of the cells of the corpus dentatum.

to the nuclei of Goll and Burdach in the medulla. Atrophic changes were also present in the direct cerebellar tract of Flechsig and the tract of Gowers (ventral and dorsal spino-cerebellar tracts). The

columns of Clarke were atrophic, and there were also some changes in the area occupied by the spino-thalamic tracts.

There was no definite atrophy of either the direct or crossed pyramidal tracts and the slight pallor in certain areas I would ascribe to

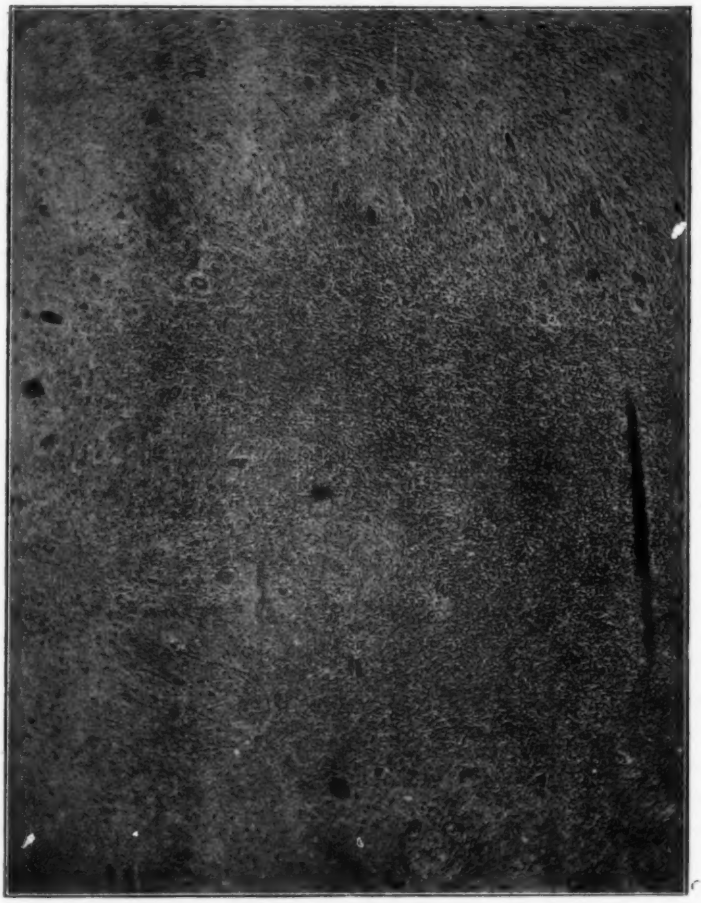


FIG. 11.—Hæmatoxylin-eosin. The corpus dentatum of dyssynergia cerebellaris myoclonica showing marked atrophy and diminution in number of the cells of the cerebellar olive. (Primary atrophy of the dentate system.)

degeneration of other non-pyramidal fibres which belong to this region.

Histological study of the brain-stem and cerebellum showed the

following conditions: There was an extension into the medulla of the system atrophies of the cord, viz., the direct spino-cerebellar tracts. The atrophy of the posterior columns apparently ceased at their ter-

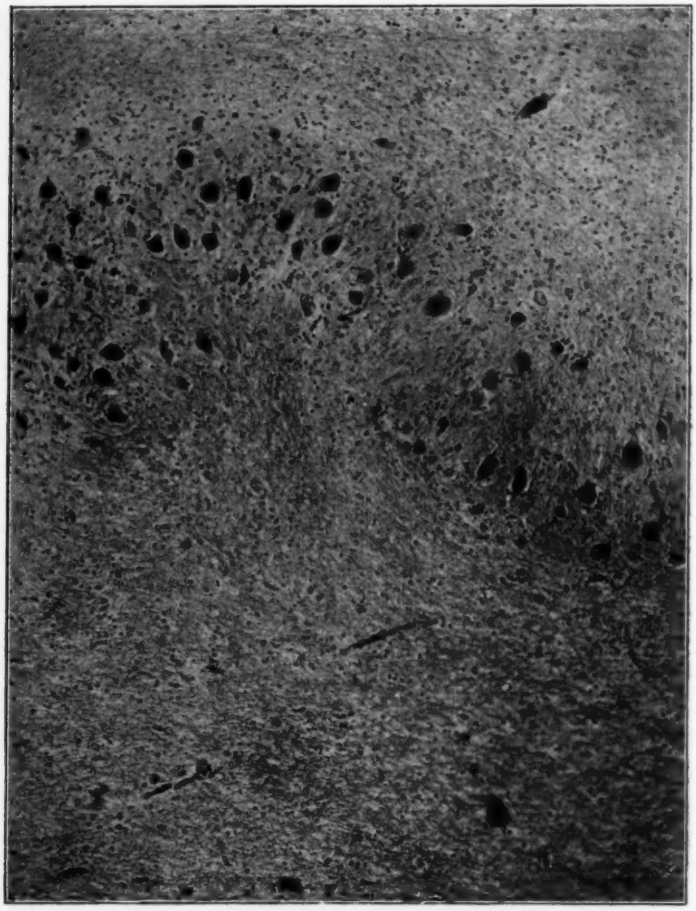


FIG. 12.—Normal control. Same magnification and stain as fig. 11. Showing normal appearance of cells of the corpus dentatum.

mination in the nuclei of Goll and Burdach, for the fibræ arcuatæ internæ were well preserved and the corpus restiforme well developed.

The other important and essential lesion was an atrophy of the

motor cells of the corpus dentatum of the cerebellum and of their efferent neurones in the superior cerebellar peduncles. There was no atrophy of the other cerebellar systems, and none of the nucleus ruber. The lesion was therefore confined to this short and very important internuncial common path which conveys the motor impulses of cerebellar origin to the spinal pathways.

The lesions were therefore both spinal and cerebellar. The spinal lesion is that common to Friedreich's ataxia; the cerebellar lesion I would regard as a special form of system disease, viz., primary atrophy of the efferent dentate system of the cerebellum.

Chapter IV.—Primary Atrophy of the Efferent Dentate System and its Relation to Dyssynergia Cerebellaris Progressiva (Progressive Cerebellar Tremor).

Anatomical Considerations.

The cerebellum is an extremely complex ganglionic structure, having massive and important connections with the spinal cord, brain stem and the cerebrum. It has an elaborate convoluted cortical structure, composed of various cellular and association layers, as well as commissural systems and peduncles. The inferior and middle peduncles are composed of afferent fibres on their way to the cerebellar cortex.

The inferior cerebellar peduncle or corpus restiforme consists of two divisions, a spinal and a bulbar. Among its constituents are the direct cerebellar tract; the anterior and posterior arcuate fibres of the nuclei of Goll and Burdach; olivo-cerebellar fibres; fibres from the nucleus lateralis and nucleus arciformis of the medulla; and the nucleo-cerebellar tracts consisting of fibres passing from the cranial nerve nuclei of the pons and medulla.

The chief constituents of the middle cerebellar peduncle or brachium pontis are: (1) the fronto-ponto-cerebellar tract; (2) the temporo-ponto-cerebellar tract; (3) the occipito-ponto-cerebellar tract; and (4) the parieto-ponto-cerebellar tract. According to Tilney and Riley [6] these fibres originate in cells of the frontal, temporal, occipital and parietal lobes respectively. They descend through the internal capsule and the cerebral peduncle to end in the cells of the pontine nuclei. From these cells arise the ponto-cerebellar fibres which decussate in the pons and pass to the cerebellar hemisphere of the opposite side.

The superior peduncle, on the other hand, is essentially efferent and

motor in function, providing the principal pathway for the distribution of cerebellar impulses to the muscular system.

This system of fibres, together with its fellow of the opposite side, undergoes a complete decussation before reaching the red nucleus of the midbrain. Its chief constituents are the cerebello-rubral tract and the cerebello-thalamic tract. It also contains a cerebello-tegmental tract.

The efferent fibres composing the superior cerebellar peduncle arise in cells of the intrinsic nuclei of the cerebellum. These are the dentate nucleus, the nucleus globosus, the embolus, and the nucleus tecti. Efferent fibres from the nucleus tecti also pass in the juxta-restiform body to the motor cranial nerve nuclei of the brain-stem (cerebello-nuclear fibres). The dentate nucleus is related to the neocerebellum or hemispheric system. Phylogenetically, it coincides in development with the cerebral cortex. The other intrinsic nuclears are older and are related to the paleocerebellum or vermian system.

All of the efferent fibres arising in cells of the intrinsic nuclei I would include under the general heading of the efferent dentate system. Those originating in the paleocerebellar nuclei may be distinguished as the *paleodentate system* and those belonging to the dentate nucleus proper as the *neodentate system.*

This would simplify our existing nomenclature for the various constituents of the efferent cerebellar system (cerebello-rubral, cerebello-thalamic, and cerebello-tegmental system), and is justified by their morphological and functional identity. The term *dentate* is chosen, as the various efferent fibre systems of the cerebellum all take their origin in the intrinsic nuclei of the cerebellum, of which the dentate is the chief constituent. This, the more recent acquisition in the phylogenetic sense, represents the neodentate system, in contra-distinction to the paleodentate system which takes its origin in the older nuclei.

Pathological Considerations.

In Chapter II I have described in some detail a group of cases characterized by symptoms of a progressive cerebellar disorder, associated with myoclonus-epilepsy.

In Chapter III a similar group of cases was reported in association with Friedreich's ataxia. In one of these cases with symptoms of a cerebello-spinal involvement, a careful histological study was made of the central nervous system, with the following results :—

The spinal cord showed the typical lesions of Friedreich's ataxia, which represented the pathological basis for the spinal symptoms of the disorder. A careful examination of the cerebellum and its connections revealed in addition an atrophy of the cells of the dentate nucleus and of the superior peduncles of the cerebellum.

The nature of the atrophic process and the strict limitation of the lesion to a special group of neurones are characteristic of a system disease. This condition I believe is a primary atrophy of the efferent dentate system of the cerebellum, and the cause of the cerebellar portion of the symptomatology.

Atrophic changes in the cells of the dentate nucleus in cases of Friedreich's ataxia have been previously described by Spiller [20], and also by Thomas and Durupt [4], in association with other lesions of this region. In the case recorded by Thomas and Durupt there were in addition to atrophy of the dentate nucleus pathological changes in the red nucleus, the fillet and the posterior corpora quadrigemina.

Like the spinal cord, the cerebellum shows a special tendency to various types of congenital defects and system diseases. These may be limited to the cerebellum but are often combined with spinal cord changes, and represent a group of cerebellum-spinal disorders. Prominent among these are the hereditary spinal ataxia of Friedreich, and the hereditary cerebellar ataxia of Marie, which may be present alone or in combination. In this disorder the lesion of the cerebellum is usually a general atrophy of the whole organ, which appears much reduced in size.

In addition to this more general type of cerebellar atrophy many more circumscribed forms are recognized, in which the lesions are limited to certain special systems of the cerebellar mechanism.

There is, for example, the olivo-ponto-cerebellar atrophy of Dejerine and Thomas [6] in which the atrophic lesions are confined to the inferior olive and its cerebellar connections, the pontine nuclei, middle peduncles of the cerebellum and the cerebellar cortex. All of the intrinsic nuclei of the cerebellum and the superior cerebellar peduncles are intact. This type is neither congenital, hereditary nor familial and appears only in later life.

Then there is a type described by Gordon Holmes [9], the olivo-cerebellar type of cerebellar atrophy. In this form there is atrophy of the cerebellar cortex, the inferior olive and the olivo-cerebellar tract. The pontine nuclei and middle peduncles, and the intrinsic nuclei and superior cerebellar peduncles are normal. This form differs from the preceding in its familial incidence.

Still another type has been added by Le Jonne and Lhermitte [15], the olivo-rubro-cerebellar atrophy. This form is characterized by atrophy of the inferior olive and olivo-cerebellar fibres, but also include the intrinsic nuclei, the superior cerebellar peduncles and the cells of the nucleus ruber. The structure of the cerebellar cortex, the pontine nuclei, and middle peduncles are intact.

The lesions in all of these types are those of simple atrophy which characterize a system-disease, and consist of primary degeneration of the parenchyma with secondary proliferation of glia.

In addition to the cerebellar system diseases there are also pure cortical types of cerebellar atrophy in which the degenerations are limited to the finer structures of cerebellar cortex (cells of Purkinje and granular layer). Such cases have been described by André-Thomas [1] and Rossi [17], and present the typical clinical picture of cerebellar disease.

At the present time we may recognize therefore the following pathological types of cerebellar atrophy. The cerebellar cortical type of André-Thomas, the olivo-ponto-cerebellar type of Dejerine and Thomas; the olivo-cerebellar type of Holmes, and the olivo-rubro-cerebellar type of Le Jonne and Lhermitte.

To these various types I would add the primary atrophy of the dentate system (figs. 9 and 11). This is a system disease characterized by atrophy of the cells of the dentate nucleus (neodentate system) and thinning of the superior cerebellar peduncles. All other important structures of the cerebellum, the cortex, the olivo-cerebellar and ponto-cerebellar systems are intact. In this group of cases the symptoms of a cerebellar disorder are much more evident in the co-ordinated movements of the extremities than in those of the trunk. The cerebellar or intention tremor which was present in greater or lesser degree in all of the cases was a characteristic symptom. The dyssynergia is, therefore, of the appendicular rather than the axial type. In this respect differing from the massive trunkal involvement of many other forms of cerebellar atrophy.

There already exists in medical literature a number of isolated facts tending to show the close connection between, what I term, the dentate system and the cerebellar or intention tremor. The relation of the intention tremor to the cerebello-rubral system was emphasized some years ago by Gordon Holmes [8], based on numerous clinical and pathological observations. Sander [18] and Touché [22] have also reported cases of intention tremor in association with lesions of the

dentate nucleus. In the experimental field, the work of Ferrier and Turner [7] on monkeys has also shown the relation of the cerebellar or intention tremor to the efferent cerebellar system. After section of the superior cerebellar peduncles these investigators observed: "Except when quite at rest, a constant tremor in the homolateral arm and leg which passed into larger oscillation on volitional effort. The limbs were the subjects of fine tremors which became amplified on exertion."

As the pathological changes in the case just recorded are limited to the neodentate system of the cerebellar mechanism, it is but natural that the more recently acquired and more highly differentiated motor activities should show the greatest degree of involvement. It is for this reason I believe that the affection is predominantly appendicular in its manifestations and the intention tremor so conspicuous a symptom, the neodentate portion of the efferent cerebellar system showing a greater vulnerability to abiotrophic degeneration.

In this respect, there is a certain similarity to another system disease which I have described in relation to the corpus striatum, viz., the primary atrophy of the efferent pallidal system [11].

This disease represents a *primary* or *essential* form of paralysis agitans. The pallidal system, like the dentate system, is a common pathway and serves for the transmission of all motor impulses from the corpus striatum. In this form of paralysis agitans the earliest involvement is usually of the extremities, which is probably due to the initial atrophy of the pallidal cells of the neostriatum (neopallidal cells). I believe, therefore, in the cerebellum as well as in the corpus striatum we must recognize the existence of an internuncial common pathway for the transmission of efferent impulses. Both systems may be the seat of abiotrophy in the sense in which this term was used by Gowers. One is the primary atrophy of the pallidal system causing the primary or essential form of paralysis agitans. The other is primary atrophy of the dentate system causing the primary or essential form of dyssynergia cerebellaris progressiva. In addition to these manifestations of primary atrophy, secondary and symptomatic forms may be recognized, caused by tumours, inflammatory and vascular lesions.

Chapter V.—The Relation of the Cerebellum to the Static or Posture System of Motility.

All investigators have recognized the important rôle played by the cerebellum in the control and regulation of motility, its influence on muscle tonus, equilibrium and the co-ordination of movement. The trend of modern opinion, under the leadership of Babinski, has been to group all cerebellar symptoms under the general heading of synergia. According to this conception, which is now generally accepted, the essential function of this important structure is the regulation of the synergies of movement, and synergia is regarded as the principal function of the cerebellum. And yet, notwithstanding this recognized rôle in the control of movement, there is much that the prevailing theories of cerebellar function have failed to explain.

In the course of previous investigations on the subject of motility I reached the conclusion that the efferent nervous system is susceptible of division into two separate components which I termed *the static and kinetic systems* [12] [13]. As this conception has an important bearing on the interpretation of cerebellar symptoms a brief outline of its chief features may be given here.

According to this view, motility is subserved by two distinct components, each represented throughout the entire efferent nervous system by separate neural mechanisms, which are physiologically and anatomically distinct. One is the movement proper, which is subserved by the *kinetic system* (motion system). The other represents that more passive form of contractility which we recognize in tonus, posture and equilibrium and is subserved by the *static system* (posture-system).

The term static is used to designate that peculiar property of the muscle fibre by which it becomes fixed in posture. In the reflex nervous system this is manifested by postural tonus and at the higher levels by various postures and attitudes.

There are many reasons for the assumption that the kinetic system is related to the anisotropic discs and the static system to the sarcoplasm of the muscle fibre, as these elements differ in structure, innervation, mode of contractility and metabolism.

There are also reasons for the assumption that the transformation of movement into posture is effected by fixation of the sarcoplasm which converts the contractile muscle fibre from a kinetic into a static mechanism.

Therefore, movement is subserved by a kinetic, and posture by a

static mechanism, the two systems supplementing one another and working together in harmony. For every movement starts from posture, is accompanied by posture, and terminates in posture, posture following movement like a shadow. At the same time, the postural mechanism exercises a stabilizing influence upon the course of movement itself.

Motility as it is observed in animal life may be divided more or less arbitrarily into three distinct groups, viz., reflex, automatic-associated, and isolated-synergic or dissociated types of movements.

All of these forms of movement are intimately associated with that other component of motility which we term posture, and the same postural groups may be recognized as in the classification of movement, viz., *reflex postures, automatic-associated postures* and *isolated-synergic types of posture* corresponding to their respective motion mechanisms. And it may be stated as a general principle that there is no form of motility from the simple reflex to the most skilled and individualistic type of movement which functions without a corresponding static or posture mechanism.

The essential integrating and correlating mechanism for the control of the static function of motility I believe to be the cerebellum. Afferent impulses from the periphery and efferent impulses from the cerebral cortex pass to this organ before their final distribution by way of the cerebello-spinal system to the posturing or static mechanism of skeletal muscles.

The older cerebellum stands in relation to what may be termed the *paleostatic system* which controls the older static or postural functions of "automatic and associated type." It takes its origin in the older nuclei of the vermis cerebelli (N. fastigii, globosus, emboliformis).

The cerebellar hemispheres, on the other hand, regulate the higher postural functions of motility through the medium of a neostatic system, which takes its origin in the cells of the dentate nucleus. The neostatic system is controlled from the cerebral cortex by special tracts which connect the various regions of the cerebral cortex (frontal, parietal, temporal and occipital) with the dorsal and ventral nuclei of the pons varolii, and thence to the opposite hemisphere of the cerebellum.

In posture, as in movement, no sharp line of demarcation can be drawn between the various types, and one group merges imperceptibly into another.

It is interesting to observe that all posture systems, *neostatic* as well as *paleostatic*, pass to the cerebellum for final integration and

co-ordination. This is in accord with the nature of the posturing mechanism and its secondary and unconscious rôle in motility. For while the higher forms of movement are initiated as conscious and voluntary processes, the corresponding postures are secondary, and follow automatically in the path of movement.

The cerebellum, then, is an organ which is engaged in the control and regulation of the static or *posture synergies* of motility. In it are represented the infracortical centres for the posturing mechanism which are under both spinal and cerebral control. I would, therefore, regard the cerebellum as a central ganglionic station for the co-ordination and control of static or posture synergies—in contradistinction to kinetic or motion synergies which are localized in their respective kinetic spheres (rolandic and striatal). The static synergies are manifestations of the sarcoplasm of the muscle fibre and the kinetic synergies of the anisotropic discs. The static or posture mechanism according to this conception is concerned in the regulation of muscle tonus, the maintenance of postures and attitudes, and is also an important factor in guiding, checking and fixation of movement.

A striking experiment cited by André-Thomas [2], may be mentioned in confirmation of this point of view. A dog whose cerebellum had been removed was unable to stand or walk. When, however, he was thrown into the water he could swim without difficulty. This peculiar phenomenon I would explain as follows: When the animal attempted to stand or walk, the posturing mechanism was essential to co-ordinated movement, to counteract the force of gravity. In a supporting fluid medium like water, however, posture synergies are of secondary importance, and as the kinetic system with its synergies are undisturbed the act of swimming takes place without difficulty.

In many respects, this theory of posture synergy also harmonizes with our modern conception of cerebellar symptomatology. In the co-ordination of all forms of movement, posture synergy is of prime importance. Loss of this control is, I believe, the cause of the scanning speech, cerebellar ataxia, intention tremor, hypermetria, adiadokokinesis and nystagmus which we observe in cerebellar disease.

In cerebellar ataxia, for example, a characteristic feature is the decomposition of movement which takes place during the execution of a co-ordinated act. When this is present it requires several disjointed movements to accomplish what is normally performed as a continuous one. The discontinuity of movement, which is one of the cardinal

symptoms of cerebellar disease, may be referred to a disorder of posture synergy which prevents the posture and motion systems from acting together in harmony. The decomposition of movement represents therefore an effort to compensate for the loss of the posturing mechanism.

Hypermetria and dysmetria of cerebellar origin may also be explained by a failure of the posture system in its function of guiding and checking movement. For in any co-ordinated act, posture formulæ are as necessary as motion formulæ and play an important rôle in giving stability and direction to movement. The checking of movement and its fixation in posture is also an essential function of the static system.

Adiadokokinesis may likewise be ascribed to a disorder of the static mechanism, the rapid succession movements which characterize this disorder being dependent upon quick fixation and sudden release of the posture mechanism.

The cerebellar or intention tremor is a particularly striking example of a disorder of the posture mechanism. When a patient presenting this symptom attempts a movement, the extremity passes into coarse ataxic oscillations which increase in intensity as the object is reached and the extremity tends to become fixed in posture. There is a coarse ataxia and tremor-like oscillation not only during the passage of the movement, but also during attempts at fixation. It is particularly at the end of the movement when the extremity tends to become fixed in posture that the intention tremor is often most active. This characteristic of the intention tremor is due to a posture asynergia, and serves to differentiate it from other forms of purely kinetic ataxia.

Cerebellar nystagmus may also be mentioned as representing a similar mechanism. Here again the movements increase when the attempt is made to fix them in posture.

Very interesting is the influence of the vestibular mechanism on the posture system of skeletal muscles. Barany has shown that all movements, trunkal and appendicular, are under a certain labyrinthine control. This is shown by the nystagmus, the pass-pointing and falling tests.

In brief then, while asynergia may be regarded as the fundamental symptom of cerebellar disease, I would limit this conception to *posture asynergia*, which is a disorder of the static or posture system of motility.

Posture asynergia may be paleostatic or neostatic, depending upon the portion of the cerebellum involved. Paleostatic asynergia is a disorder of lower forms of posture of automatic-associated type, and is largely trunkal or axial in distribution. Neostatic asynergia, on the other hand, is a disorder of the higher forms of posture, of isolated synergic type and is essentially appendicular in distribution. One is a spinal, the other a cerebral form of asynergia.

In the system disease of the cerebellum described in the previous chapter—"Primary Atrophy of the Dentate System"—the atrophic changes were limited to the cells of the corpus dentatum (neodentate system). This is a system which is related to the neocerebellum and its recently acquired functions. For this reason the cerebellar symptoms of this disorder are predominantly appendicular in distribution, the extremities representing in the realm of posture, as in movement, the highest degree of development.

Chapter VI.—Concluding Remarks.

In the previous chapters, under the title, "Dyssynergia Cerebellaris Myoclonica," I have described what appears to be a well-defined type of nervous disease, presenting the clinical picture of a progressive cerebellar disorder in association with myoclonus-epilepsy. Four of the cases were sporadic and no history could be elicited of any hereditary or familial tendency to either cerebellar disease or myoclonus-epilepsy.

The cerebellar disturbance affected more especially the finer movements of the extremities and the dyssynergia was predominantly appendicular in distribution. The cerebellar or intention tremor was a marked feature of all the more advanced cases, in this respect resembling the clinical group which I described some years ago under the title "Dyssynergia Cerebellaris Progressiva," or "Progressive Cerebellar Tremor."

The myoclonus-epilepsy, in its general manifestations, was very similar to the disorder as described by Unverricht [23]. The familial incidence, however, which is one of the chief characteristics of the Unverricht type, was lacking, excepting in the group associated with Friedreich's ataxia. The myoclonus-epilepsy, therefore, resembles in some respects the type of the disorder described by Rabot [16]. This is a sporadic form, beginning somewhat later in life than the Unverricht type and in which the myoclonus is intermittent in character and less

progressive in its evolution. Fundamentally, however, both types are very similar and are united by transition forms.

The relation of the cerebellar disorder to myoclonus-epilepsy in the group of cases which I have described is quite obscure and in the present state of our knowledge but little light can be thrown on this question. It is quite possible that the combination is only accidental and represents the association of two independent nervous disorders in a predisposed individual. Such combinations in the realm of neuropathology are by no means uncommon. Nevertheless, I think that one should not be too hasty in concluding that here is a mere combination of separate clinical entities. We know so little of the underlying cause and pathology of myoclonus and its relation to the various structures of the central nervous system that the possibility of a form related to the static or posture system should be considered. It is conceivable, for example, that sudden breaks in the continuity of postural control or synergy might express themselves in terms of compensatory movements of a myoclonic type.

Interesting in this connection are the peculiar epileptiform attacks which occurred in two of the cases, which I have termed static seizures. These attacks are characterized by sudden loss of postural control without convulsive manifestation and are accompanied by slight transitory obscuration of consciousness. The patient without warning plunges suddenly to the ground, apparently from a sudden cessation of postural control. These attacks are so sudden and the fall so immediate that injury is very likely to ensue. There are no muscular spasms and the patient rises almost immediately. The blurring of consciousness is often only momentary; indeed in some of the attacks consciousness is apparently unaffected.

In addition to the clinical type, termed dyssynergia cerebellaris myoclonica, a similar complex of symptoms was observed in another group of cases associated with the spinal symptoms of Friedreich's ataxia. These, however, were of the family type, two cases occurring in one family. Mention was also made of an observation recorded by Boschi showing that such combinations of symptoms, while rare, are not unknown in medical literature.

In one case, belonging to this group, a careful histological study was made of the central nervous system, which revealed the typical spinal lesions of Friedreich's ataxia, in conjunction with a peculiar type of system atrophy of the cerebellum, which I termed primary atrophy of the dentate system. The atrophic changes involved only the cells of the

corpus dentatum and the fibres of the superior cerebellar peduncles. The pathological changes were therefore limited to the cells and fibres of a special system of neurones, which because of their relation to the dentate nucleus was termed the dentate system. All other structures of the cerebellum were normal.

To the other recognized system atrophies of the cerebellum, viz. : the cerebellar cortical atrophy, the olivo-ponto-cerebellar atrophy; the olivo-cerebellar atrophy and the olivo-rubro-cerebellar atrophy, I would add the atrophy of the dentate system, which is a primary atrophy of its chief efferent system. The association of this form of cerebellar atrophy with Friedreich's ataxia is not surprising in view of the frequency of cerebello-spinal combinations.

The cerebellar portion of the symptomatology of dyssynergia cerebellaris myoclonica I would refer to the atrophy of the efferent dentate system. In the present state of our knowledge it is difficult, if not impossible, to refer general cerebellar symptoms to special parts of the cerebellar mechanism. And it is generally held that the various types of cerebellar atrophies mentioned above cannot be distinguished with certainty on the basis of their cerebellar symptomatology alone. In other words, asynergia is the characteristic symptom of cerebellar disease, no matter which portion of the cerebellar mechanism is involved.

It is, therefore, with some hesitation that I suggest the early appearance and predominance of appendicular asynergia and especially the cerebellar tremor as perhaps characteristic of this type of cerebellar disease (primary atrophy of the dentate system) and yet these were the characteristic features of the clinical type under consideration.

Some years ago I described a primary system disease of the corpus striatum which was associated with the symptomatology of paralysis agitans. The lesions consisted of atrophy of the large motor or pallidal cells of the corpus striatum, and of their efferent fibres in the ansa system. The disorder was called primary atrophy of the pallidal system. This I regarded as the primary and essential form of paralysis agitans and all other clinical manifestations of the disease as secondary or symptomatic in nature due to vascular, inflammatory and neoplastic lesions.

A similar differentiation may be made in the case of the cerebellum and its efferent pathway, the dentate system. This may be the seat of a primary atrophic process producing the characteristics of a cerebellar disorder (primary atrophy of the dentate system). This system may

also be the seat of secondary or symptomatic involvement from tumours, inflammations and vascular lesions. The intention tremor of multiple sclerosis is a symptomatic manifestation of involvement of this system.

Primary atrophy of the pallidal system is, therefore, a disorder of a kinetic mechanism, giving rise to the symptomatology of paralysis agitans. Primary atrophy of the dentate system is a disorder of a static mechanism, giving rise to the symptomatology of dyssynergia cerebellaris.

In the final chapter I presented my views on the functions of the cerebellum and its relation to the static system of motility. According to this conception the function of the cerebellum is the control of the posture synergies of movement. Motility is subserved by two components throughout the whole of the efferent system, a kinetic or motion system and a static or posture system, both working together in harmony.

Motility, operating through its contractile mechanism, is therefore a compound of movements and postures. Movement is the active component which propels the organism or parts of the organism in space, the peripheral contractile mechanism of which is represented in the anisotropic discs of the striated muscle fibre. Posture is that more passive component which is represented in tonus and attitude. It tends to counteract the forces of gravity and the effects of atmospheric pressure and maintains the organism in a state of static equilibrium. The peripheral organ of this mechanism is the sarcoplasm of the muscle fibre. This system also plays an important rôle in the guiding, checking and fixation of movement. The various symptoms of cerebellar disease which are grouped under the general heading of asynergia may be attributed to a disorder of this mechanism.

BIBLIOGRAPHY.

[1] André-Thomas. "Atrophie lamellaire des cellules de Purkinje," *Revue Neurol.*, 1905, **13**, 917.

[2] *Idem.* "Cerebellar Functions," *Journ. of Nerv. and Ment. Dis.*, Monograph Series, New York, 1912, Chap. 11.

[3] *Idem.* "Le cervelet" (Case 5), Paris, 1897.

[4] André-Thomas and Durupt. "Examen du névraxe dans un cas de maladie de Friedreich," *Revue Neurol.*, 1912, **24**, 317.

[5] Boschi, Gaetano. "Ataxie héréditaire avec paramyoclonus multiplex; type Unverricht," *Journ. de Neurol.*, 1913, **18**, 141.

[6] Dejerine and Thomas. "L'atrophie olivo-ponto-cérébelleuse," *Nouv. iconog. de la Salpêtrière*, 1900, **13**, 330.

[7] Ferrier and Turner. "A Record of Experiments illustrative of the Symptomatology and Degenerations following Lesions of the Cerebellum and its Peduncles in Monkeys," *Phil. Trans. Roy. Soc. of Lond.*, 1894, **184**, (B), 719.

[8] HOLMES, GORDON. "Certain Tremors in Organic Cerebral Lesions," *Brain*, 1904, **27**, 327.
[9] *Idem.* "A Form of Familial Degeneration of the Cerebellum," *Brain*, 1907, **30**, 466.
[10] HUNT, RAMSAY. "Dyssynergia Cerebellaris Progressiva: A Chronic Progressive Form of Cerebellar Tremor," *Brain*, 1914-15, **37**, 247.
[11] *Idem.* "Primary Atrophy of the Pallidal System: A Contribution to the Functions of the Corpus Striatum," *Brain*, 1917, **40**, 58, and *Arch. Int. Med.*, 1918, **22**, 647.
[12] *Idem.* "The Static and Kinetic Systems of Motility," *Arch. of Neur. and Psych.*, 1920, **4**, 353.
[13] *Idem.* "The Static or Posture System and its Relation to Postural Hypertonic State of the Skeletal Muscles," *Neurol. Bull.*, 1921, **3**, 207.
[14] JENDRASSIK. "Hereditäre Krankheiten," Lewandowsky's "Handbuch der Neurol.," **2**, 394.
[15] LEJONNE and LHERMITTE. "Atrophie olivo-rubro-cérébelleuse," *Nouv. iconog. de la Salpêtrière*, 1909, **22**, 603.
[16] RABOT. "De la myoclonie épileptique," *Thèse de Paris*, 1899.
[17] ROSSI. "Atrophie primitive parenchymateuse du cervelet à localisation corticale," *Nouv. iconog. de la Salpêtrière*, 1907, **20**, 66.
[18] SANDER. "Ein pathologisch-anatomischer Beitrag zur Function des Kleinhirns," *Deut. Zeit. f. Nervenheilk.*, 1898, **12**, 363.
[19] SIOLI. "Ueber histologischem Befund bei familiärer Myoklonus-Epilepsie," *Arch. f. Psychiat.*, 1913, **51**, 30.
[20] SPILLER. "Friedreich's Ataxia," *Journ. of Nerv. and Ment. Dis.*, 1910, **37**, 411.
[21] TILNEY and RILEY. "The Form and Functions of the Central Nervous System," 1921, 456.
[22] TOUCHE. "Deux cas de ramollissement du cervelet," *Revue Neurol.*, 1900, **8**, 149.
[23] UNVERRICHT. "Ueber familiäre Myoclonie," *Deut. Zeit. f. Nervenheilk.*, 1895, **7**, 33.

ON DISORDERS OF MOVEMENT RESULTING FROM LOSS OF POSTURAL TONE, WITH SPECIAL REFERENCE TO CEREBELLAR ATAXY.

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CHAPTER I.—INTRODUCTION.

CHAPTER II.—THE REACTIONS OF THE ATONIC MUSCLE.

(1) The reflex contraction of the de-afferented muscle.
(2) The reflex contraction of the atonic muscle of the spinal animal.
(3) The nature of the tonic reactions lost to the atonic muscle.
(4) Atonia and reflex coordination.
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CHAPTER III.—THE CEREBELLAR SYMPTOM-COMPLEX.

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CHAPTER I.—INTRODUCTION.

THE problems presented by cerebellar lesions, whether experimentally produced or the result of disease in man, have been the subject of much earnest research ever since Flourens published his classic observations a century ago. Perhaps no part of the nervous system has been more intensively studied than the cerebellum, and yet the final analysis of its symptom-complex and the determination of its functions have both so far eluded us.

When we consider the complexity of the physiological problem involved, it is perhaps not surprising that we have not been able to determine the functions of this organ, but it is more noteworthy that a symptom-complex, so closely and so repeatedly studied, should still be so variously described and so diversely interpreted.

The student of the subject who seeks for some underlying basis of agreement in the various accounts of the symptom-complex contained in the vast literature of the cerebellum, is as much dismayed by the redundant and elaborate terminology employed as by the various and conflicting senses in which one and the same term is used by different

writers. Indeed, he does not discover that this oppressive and confusing vocabulary conceals a very large measure of agreement as to the facts of observation, until he discards it. Although the impression of confusion and lack of precision obtained from the literature is in part the fruit of a terminology loosely conceived, and still more loosely employed, it has its roots yet deeper in the fact that until recently the state of knowledge did not permit of any precise or accurate formulation of the essential elements of the problem. When so little was known of the nervous processes underlying the co-ordination of movement, and while it was still thought that the cerebellum was concerned with the postural aspects of sensation, a certain vagueness of thought was inevitable. When we recall the severe handicaps under which the pioneer workers laboured in this respect, we cannot but admire the scope and accuracy of their observations.

However, within the past twenty-five years, the classic researches of Sherrington and their development by Graham Brown, Magnus and others, have placed at our disposal a beautiful series of minute analyses of the modes of nervous activity, particularly in respect of the co-ordination of movement and of posture. In consequence, the modern student of the cerebellum has an immeasurable advantage over the investigator of the last century. We can approach the problem with more precise conceptions of the processes concerned in the co-ordination of movement, and with a scientific nomenclature in which to express our conclusions. Moreover, it is now possible to re-survey the earlier observations and, in the light of recently acquired knowledge, to explain much that was formerly obscure.

It is remarkable that, so far, no such re-survey has been attempted, and even the most recent experimental and clinical studies of the cerebellum have not laid this fundamentally important knowledge under contribution. This must be the excuse for the present attempt to examine what we believe to be the fundamental cerebellar defect-symptom from a point of view not yet adequately developed in this connection. It is probable that adherence to what are now obsolete conceptions of muscle tone, and the tendency to regard each particular expression of one and the same disorder of co-ordination as an elementary defect-symptom, are among the principal reasons that recent studies have not carried our knowledge beyond the point at which it stood when Luciani published his observations over thirty years ago. If we take the descriptions by various authorities of the cerebellar symptom-complex in animals and man, and, abandoning the nomen-

clature employed, tabulate in simple descriptive terms the several disorders of muscular contraction and of movement recorded, we can construct such a classification as the following :—

(*I*) *Abnormalities of resting muscle.*—Loss of tone.

(*II*) *Abnormalities of muscular contraction.*—(1) Excessive range and force of contraction ; (2) intermittent and unsteady contraction ; (3) premature relaxation ; (4) extreme readiness of fatigue.

(*III*) *Abnormalities of voluntary movement.* — Faulty functional combination of the muscles engaged in a voluntary movement.

(*IV*) *The effects of voluntary efforts at correction.*—Carried out by a musculature functionally defective in the above senses.

It is apparent at once that this classification cannot be regarded as a final analysis of cerebellar ataxy. For example, the disorders included under heading (*II*) must of necessity impair the perfect functional combination, or synergy, of muscles in a co-ordinated movement, especially in complex purposive movement-combinations. Therefore, asynergia, as the loss of this combination is called, cannot be regarded as an elementary defect-symptom, it must be considered as a manifestation of underlying disorders of muscular contraction. Further, it remains to be determined whether the disorders of muscular contraction are themselves elementary defects, or simply the result of loss of tone. This brings us to the point from which the present study begins, namely, the consideration of the part played by atonia in the production of the cerebellar symptom-complex. As we may readily see from a comparative study [19] of the various descriptions of the cerebellar symptom-complex, a considerable degree of conflict and uncertainty obtains in the use of the term " atonia." It has been variously understood by different writers on the cerebellum, and by none of them has it been used in the sense now adopted by physiologists, namely, as a diminution or loss of postural tone.

Since our conception of muscle tone must govern our use of the term " atonia," a precise definition of what we understand by tone is an essential preliminary to the study we propose. We may say that the modern physiological conception in this respect has scarcely influenced clinical thought as yet. For the clinician, muscle tone is that state of active tension characteristic of living muscle in normal connection with the nervous system, which is felt as a slight but definite resistance to palpation and to passive stretching of the muscle. Conversely, atonia has been taken to mean the loss of this active tension so that the muscle is flaccid, unduly extensible and presents no resistance to passive

stretching. These are the conceptions of tone and atonia employed in the literature on the physiology and symptomatology of the cerebellum. Even with these very restricted interpretations, there is no general agreement as to the importance or constancy of atonia as a cerebellar symptom. Thus, while Luciani [5] and Gordon Holmes [3] accept it as one of the fundamental defect symptoms, Babinski [2] and André-Thomas [1] think it rare and of no importance. Further, the effects of atonia on muscular contraction and on co-ordination of movement are differently interpreted even by those who regard it as an essential symptom. Finally, André-Thomas, who does not employ the term at all, somewhat paradoxically develops a conception of loss of tonic postural nervous activity as a fundamental cerebellar defect symptom, which more truly corresponds to the physiological sense of atonia than any interpretation to be found elsewhere in the literature of the subject.

The sense in which we shall understand muscle tone here is that which Sherrington has so clearly defined [15]. Muscle tone is a purposive reflex reaction, a proprioceptive reflex whose source is in the tonic muscle itself. From his analyses of tone in the decerebrate animal, Sherrington has shown that its physiological purpose is nothing less than the maintenance of posture. Under these conditions, it has a selective incidence in the extensor muscles of the neck, trunk and limbs, that is, in the muscles which maintain the animal in its normal attitude against the force of gravity, the "anti-gravity" muscles. Further, like other reflex reactions employing muscles, it is governed by reciprocal innervation, so that tone is inhibited in the antagonists of the tonic muscles. The hypertonus of the decerebrate animal can therefore be described as "reflex standing." Recently, Magnus has carried out a long series of experimental analyses of the reflex reactions by which tone is regulated and adapted to the varying attitudes imposed upon the animal under experimental conditions. Moreover, he has found that these reactions also obtain, and are the sum total of the reactions occurring in the intact animal in its adoption of all the attitudes of which it is capable. We may conclude, therefore, that muscle tone is the basis of posture and of co-ordinated postural adaptation. This is the only sense in which the term "tone" is employed in this paper, and by atonia we shall understand a loss or impairment of the tonic postural reactions of the normal musculature. If we are to determine the significance of such loss of postural tone in the production of cerebellar ataxy it is by an approach from the physiological standpoint alone that we can hope to reach a satisfactory solution. Therefore, we must begin

with a brief review of the peculiarities of the muscle rendered atonic by experimental procedures in animals. There are two preparations which have been employed by Sherrington in this connection, they are the "decerebrate" and the "spinal" animals. In the former the brain-stem is transected between the levels of the anterior and posterior colliculi. Following this procedure, the extensor muscles of the animal enter into a state of heightened tone. In such an animal preparation a muscle, commonly the extensor of the knee, vasto-crureus, is rendered atonic by "de-afferentation," that is, by section of its afferent nerve supply at the level of the posterior roots. From the reactions of this muscle, the results of loss of postural tone upon reflex reaction can then be investigated. The "spinal" animal is one in which the spinal cord has been transected at some level between the medulla and the lower thoracic segments. The reflex arc of postural tone has its reflex centres in the brain-stem, and therefore such a section cuts the arc and abolishes tone in the muscles innervated from below the level of the transection. However, as we shall see, the atonia produced in this way differs somewhat in degree from that produced by de-afferentation of a muscle in the decerebrate animal.

From a study of the observations of Sherrington under these experimental conditions, the conclusion is reached that *all the component symptoms of cerebellar ataxy are nothing more than manifestations of loss or impairment of postural tone, in other words, atonia is the fundamental cerebellar defect-symptom, all other symptoms being merely particular manifestations of this one defect.*

Chapter II.—The Reactions of the Atonic Muscle.

There can be no doubt that loss of postural tone involves a serious disturbance of balance in the reflex centres concerned in the co-ordination of movement. In the following brief description of the effects of loss of tone upon reflex co-ordination, as Sherrington has revealed them to us, we may hope to reach more accurate conclusions than is possible from clinical observations alone.

(i) *The reflex contraction of the de-afferented muscle.*—Briefly stated, the atonic muscle under these conditions is unduly excitable, contracts more amply than normal and relaxes at once upon cessation of the reflex stimulus. These peculiarities may be considered in detail. The threshold of stimulation, both for excitation and for inhibition, is commonly lowered. Occasionally, however, the threshold may be

normal or even supernormal. The latency of toneless muscle is not altered. The characters of the response are very striking. Contraction develops rapidly and rushes to a maximum, which exceeds the normal, "as though its momentum were less controlled than normal" [11] [13] [14]. Not only is contraction excessive in range, but it is very unsteadily maintained and dies down suddenly as soon as it reaches its height. It does not outlive the exciting stimulus, that is, there is no "after-discharge." A striking result of the last feature is the coarsely clonic or intermittent character of the response to serial stimuli. The normal fusion of component contractions is absent or much impaired [11]. The post-inhibitory exaltation, which normally gives rise to "rebound" contraction, is absent, or is tremulous, ill-maintained and feeble [13]. Fatigue sets in rapidly, so that the response soon wanes, especially in weak contractions [14]. The de-afferented atonic muscle is very susceptible to inhibition, and under this the contraction yields as irregularly as it responds to excitation [14].

(ii) *The reflex contraction of the atonic muscle of the spinal animal.* —The features we have described are equally characteristic of the response obtained from the atonic muscles of the spinal animal, though here they are present to a less extreme degree. In the hind limbs of such a spinal animal several other results of loss of postural tone may be observed. Stimuli, which in the tonic muscle of the decerebrate animal give rise to maintained tonic responses, now evoke alternating movements [13]. Thus, pinching the foot elicits tonic homolateral flexion and crossed extension reflexes in the decerebrate animal, but in the spinal animal clonic alternating movements of flexion and extension, the response opening with homolateral flexion and crossed extension. Similarly, pinching the skin of the perineum in the former results in tonic bilateral extension of the hind limbs, but in free stepping movements in the latter [13]. Finally, the spread of reflex response to regions of musculature remote from that stimulated is much more facile in the spinal than in the decerebrate animal.

(iii) *The nature of the tonic reactions lost to the atonic muscle.*—To understand these phenomena we must have a clear idea of the characters and purpose of the tonic reactions which are absent in the de-afferented muscle and grossly impaired in the muscles of the spinal animal. When a reflex response is elicited in the tonic musculature of the decerebrate animal, the muscular contraction rises rapidly to its height and then declines slowly and often incompletely. The muscle tends

to remain shortened at the new length imposed upon it by the contraction. This "shortening reaction," as Sherrington [11] has called it, is a proprioceptive reflex produced by the initial reflex contraction of the muscle. Like other proprioceptive reactions it is tonic and in effect reinforces the original exteroceptive reflex. Conversely, if the muscle be actively inhibited by some exteroceptive reflex stimulus, the new and increased length reached is maintained by a secondary proprioceptive inhibitory reaction, the "lengthening reaction." The "shortening reaction" is responsible for the tonic "after-discharge" and ensures a steadily maintained contraction in response to serially repeated stimuli. Moreover, after inhibition of tonus, just as after inhibition of active movement, there is a rebound, or post-inhibitory exaltation of tone, which ensures a return to the initial posture broken into by the movement. Thus, these two tonic reactions act as adjuvant and as compensatory reflexes, steadying and maintaining the original contraction and ensuring a return to equilibrium when this has ceased. They give a quality of "plasticity" to muscle tone in virtue of which posture is maintained in the varying attitudes adopted by the living animal.

The effect of the shortening reaction on reflex movement is best and most simply seen in the knee-jerk. As Sherrington [15] and Viets [17] have both shown, there are two elements in the normal knee-jerk, an initial twitch produced by the tap on the tendon, and superimposed upon this a secondary proprioceptive tonic after-discharge or shortening reaction. In a myogram it is seen as a gradual and incomplete fall of the limb. This shortening reaction is entirely absent in the knee-jerk obtained from the atonic muscle, in which the relaxation is sudden and complete.

(iv) *Atonia and reflex co-ordination.*—If the reflex contraction of a muscle is disordered by loss of tone in the manner we have described, it follows that the reflex co-ordination of movement must suffer even more. In the case of the simple reflex elicited by a chosen stimulus we have two groups of muscles involved, agonists and antagonists. Under reciprocal innervation, when the former group is excited to contract, the latter is inhibited. The atonic muscle behaves as irregularly under inhibition as when contracting. So that side by side with excessive amplitude of movement, abruptness of onset and cessation, fatigability, irregular yielding to inhibition, and, in the serial responses, a clonic intermission of the contraction wave, we have in addition a negative side of all these defects present in the antagonist group. But the simple reflex, employing reciprocal innervation in an otherwise quiescent muscu-

lature, is an abstraction, an artificial state of affairs never reproduced during life [14]. Here conflicting stimuli pour continuously upon the surface of the organism and compete for possession of the different muscular mechanisms. At a given moment, therefore, any reflex centre is under a twofold influence, one tending to excite the muscles governed by it to contraction, the other to inhibit them, or, at least, to employ them in a reaction of different character and purpose. In the normal organism the result is not chaos but perfect co-ordination. Sherrington has called the process by which this is achieved *double reciprocal innervation* [12] [14]. When two conflicting stimuli struggle for possession of a final common path, the resulting reaction depends upon their relative intensities. If there be considerable disparity, one displaces the other which is suppresed, but if their intensities be relatively equal the result is an algebraical summation of the effects. The muscle contracts, but less powerfully than it would have done under a single exciting stimulus, and its antagonist is inhibited, but less completely than it would otherwise have been. Therefore, each muscle is under a twofold influence, it is at once excited and inhibited. In the agonist the former effect predominates, in the antagonist the latter. Therefore, both muscles of an antagonistic pair are in movement or in tonic contraction simultaneously. But under double reciprocal innervation, phasic or tonic contraction does not increase simultaneously in both. As it waxes in one it wanes in the other, except when both start from quiescence, when contraction is stronger in the agonist than in the antagonist. This combination of excitation and inhibition is a fundamental factor in the grading of reflex contraction.

The atonic muscle with its loss of balance and stability in reflex reaction must, under these complex conditions, give rise to still more marked disorders of co-ordination. Moreover, in the normal individual voluntary movement involves the breaking up of the simple reflex movements into combinations and sequences of almost infinite variety and complexity [4]. Vitiating all these complex activities in direct proportion to their complexity is the uncontrolled, abrupt and unsteady character of reflex response occurring in an atonic musculature. We see that tone is intimately interwoven with movement in the processes of perfect co-ordination. To use Jackson's expressive phrase, there is "a perfect co-operation of antagonism" between the phasic and tonic elements in the co-ordination of movement. How intimate this association must be, and how essential its maintenance, is apparent when we remember that all our movements demand the finest postural adapta-

tion. The spinal animal can step, but it cannot walk because it is unable to stand. The lecturer who writes on a blackboard maintains his arm in the extended and elevated position while his forearm and hand muscles execute the movements of writing. This postural adaptation is achieved by the reflex modification of tone. If his muscles be atonic, he has consciously and by voluntary direction to hold the arm extended by a series of movements each of which subsides immediately unless it be repeated. The contractions underlying this voluntary replacement of what is normally a subconscious reflex process will have all the characters of an atonic musculature; they will be excessive in range and force, subject to premature relaxation, to fatigue and to intercurrent inhibition. In the maintenance of the erect posture and in movements of locomotion where postural reactions play an even more obviously important part, it is inevitable that there should be the gravest disorders of co-ordination, and therefore of the maintenance of equilibrium.

(v) *The spinal component of tone.*—We cannot leave the discussion of atonia without reference to the significance of the spinal component of tone. Tone is observed in the muscles of the spinal animal after recovery from "spinal shock." For various reasons it is more readily studied in the dog than in the cat. It is never observed in the spinal monkey [9]. The immediate result of spinal section in the lower thoracic region in the cat is to produce a flaccid paraplegia. A brisk knee-jerk is obtainable immediately after recovery from the anæsthetic. By the next day there is usually appreciable tone in the hip flexors, and very shortly in the adductors of the thigh. At first the extensors remain flaccid, but, even in these muscles, in four or five days transient accesses of tone may be observed. In every instance, and whatever group of muscles is in question, this spinal tone is peculiarly fugitive and uncertain of development. It is liable to sudden and complete lapses. A definite shortening reaction is rarely obtained, and, in short, spinal tone lacks plasticity and is most ineffectual as a postural reflex. Sherrington [11] finds that the spinal dog can be made to stand for a few seconds, but here, as in the cat, tone is apt to disappear suddenly and the animal collapses. Therefore, while passive movement may indicate the presence of tone in the muscles under these conditions, yet in the strict sense postural tone may be considered to be absent. This is an observation not without significance in relation to the atonia of cerebellar lesions, and will be discussed later.

While it is found that in the decerebrate animal postural tone has a selective incidence in the extensor group of muscles and is not readily discoverable in flexors, we must remember that decerebrate rigidity is an exaggerated tone. Since reciprocal innervation obtains for tone, it follows that in the decerebrate animal flexor tone is unduly inhibited. We might expect the distribution of tone in the intact animal to show less inequality in this respect, though all the evidence indicates that the extensors form the more important group where tone is concerned. This is readily comprehensible when we recall that they are responsible for the maintenance of the normal attitude of standing. The observations of Magnus are of great interest in this connection [6] [7]. He finds, in confirmation of the earlier work of Thiele [16], that decerebrate rigidity does not appear until the plane of transection of the brain-stem passes caudal to the anterior colliculi. An animal (rabbit, cat, dog or ape) in which the plane of transection passes immediately anterior to the mid-brain does not show decerebrate rigidity, its tone has what Magnus calls a "normal distribution." Such a mid-brain animal can not only stand, it can walk and jump and, if overturned, can right itself again. All its postural mechanisms are intact. In other words, the reflex arcs necessary to the maintenance of tone, and the reflex mechanisms by which it is regulated in the interests of co-ordination, are all intact.

When the transection lies sufficiently caudal to produce decerebrate rigidity some of these regulating mechanisms have been removed and simple tone, released from control, remains. In the de-afferented muscle of the decerebrate animal and in the atonia of the spinal animal the arcs concerned in the maintenance and the regulation of tone have both been cut out.

(vi) *The muscle rendered atonic experimentally, compared with the "ataxic" muscle of cerebellar lesions.*—At first sight there might appear to be one grave objection to the comparison of such relatively artificial preparations as muscles rendered atonic by the experimental procedures indicated with the "ataxic" muscle of a case of cerebellar disease or injury. It is true that the de-afferented muscle of the decerebrate preparation has been deprived of both sensory and non-sensory afferent fibres, while the affected muscle in cerebellar disease has its sensory path intact. But in the decerebrate animal the sensory path has already been interrupted by the removal of the cerebral hemispheres, and yet tone and all tonic reactions are intact. Therefore the absence of sensory afferent fibres does not introduce a complicating factor.

The maintenance of tone is a function of the non-sensory afferent group, and it is to the physiological disturbances in the central connections of this group that the "ataxy" of cerebellar lesions is due.

The condition of the muscles of the spinal animal is somewhat different. Here also the sensory path is destroyed together with the long reflex arc of postural tone. The local spinal arc, however, is intact and hence the purely spinal, phasic component of co-ordination remains.

In these circumstances, some tone, it is true, does return to the muscles, but it has little plasticity or postural value, is inconstant, liable to sudden complete lapses and is not comparable with the extensor muscle tone of the decerebrate preparation. Hence the relatively atonic muscle of the spinal animal shows the defects seen in the de-afferented muscle, though to a less complete degree.

From another point of view there appears at first sight a difference between the activities of the atonic muscle in the experimental animal and those of the "ataxic" muscle of the subject of a cerebellar lesion. The former is capable of reflex responses alone, while the ataxy of cerebellar lesions is manifested in voluntary activities. From what we know of the functions of the cerebral motor cortex (Leyton and Sherrington) it appears probable that the actual processes of co-ordination are a function of the lower reflex levels of the nervous system, and voluntary movement is but the activation of these reflex mechanisms for the production of an infinite variety of complex movement combinations and sequences. The large and relatively simple motor reactions of the decerebrate or spinal animal are the material from which, by a process of analysis and synthesis, the cerebral cortex elaborates the skilled and finely adapted movements of the normal individual. In the subject of a cerebellar lesion, just as in the experimental animals we have been considering, it is the activity of the reflex levels which is interfered with, so that we may conclude that the presence of volitional activity in the case of purely cerebellar lesions does not invalidate the comparison we are about to make between the disorders of co-ordination produced by loss of postural tone aud the motor disorders which we include under the heading of cerebellar ataxy.

Chapter III.—The Cerebellar Symptom-Complex.

We are now in a position to review the cerebellar symptom-complex in the light of the experimental observations we have described. So apparent is the identity of many of the symptoms of cerebellar lesions with the characteristic reactions of the atonic muscle that no lengthy consideration is required. If we take the disorders included under heading (*II*) of the table on p. 541, we see that they are all disorders which occur when postural tone is abolished. We have already discussed the clinical conception of atonia. Its limitations are sufficient to explain how it is that "atonia" has not been regarded as of more importance by the clinician. It is true that Luciani concluded that atonia, asthenia and astasia were but different expressions of a single defect, and that his definition of tone includes phenomena which subsequent experimental research has shown to be manifestations of loss of postural tone. We cannot but regret that physiological knowledge of the processes concerned in the co-ordination of movement was not more advanced at the period of Luciani's investigation. For so accurate are his observations and so penetrating his conclusions that could he have reviewed them from the standpoint of modern knowledge of the physiology of the nervous system, we might well have been within sight of a satisfactory theory of cerebellar functions.

There are authorities who deny the occurrence of atonia, in the clinical sense, as a cerebellar symptom, or who believe that muscle tone may be restored before perfect co-ordination returns. From what has been said of the spinal component of tone we can readily understand that some recovery from the profound muscular flaccidity of a recent acute cerebellar lesion may well occur while ataxy is still observed, or that in other instances no complete flaccidity on palpation or on stretching may be seen. Yet this does not necessarily indicate that postural tone is present in the cases in question. As we have seen, the tests employed for the estimation of tone, as this is understood clinically, are not competent to decide this point. However, if we accept the hypothesis now put forward that cerebellar ataxy is itself the direct and inevitable result of postural atonia, we shall no longer be restricted to these inadequate standards, but shall have every test for cerebellar ataxy as a criterion of postural tone.

While it is scarcely necessary to go *seriatim* through all the disorders of voluntary movement observed in the subjects of cerebellar lesions, we may briefly take one or two of the most characteristic ones

and consider them as possible results of postural atonia. Let us take, for example, the ready fatigue, the excessive range of movement ("dysmetria") and the oscillations or jerky variations of sustained movements ("discontinuity of movement"). They are found to be, without exception, inevitable results of loss of postural tone.

As Luciani pointed out, we cannot exclude the complicating effect of voluntary efforts at compensation and correction, themselves carried out in an atonic musculature. They must aggravate the disorder of movement in voluntary motor activities, but in the ultimate analysis atonia or loss of postural tone is an adequate explanation of all the manifestations of cerebellar ataxy. Finally, the graphic records of the knee-jerk obtained in cases of acute cerebellar lesion by Gordon Holmes show an ample twitch contraction without shortening reaction, a typical atonic reflex response. André-Thomas has also described this type of knee-jerk as characteristic of cerebellar lesions.

A further point remains for consideration. Luciani observed that after ablation of the cerebellum, and at a stage when it could not stand, a dog thrown into water was able to swim without any defect of co-ordination. From this he concluded that inco-ordination, in the strict sense of the term, did not occur as a result of cerebellar lesions. The essential defect was an asthenia in virtue of which the animal was not able to support its weight, and the water, by overcoming this difficulty, rendered perfect harmony of movement possible. This remarkable observation does not appear to have attracted the attention it merits. In the light of the present hypothesis it assumes considerable significance, and may be taken to afford some confirmation of the point of view presented in this paper. Movement in such a resistant medium as water must, though imperfectly, control many of the most characteristic results of loss of postural tone; for example, the suddenness of contraction and of relaxation, the excessive amplitude and force of contraction, and the oscillations resulting from loss of postural fixation. Indeed, we could scarcely devise an experiment more calculated to demonstrate the fundamentally important rôle of atonia in the production of cerebellar ataxy.

In conclusion, the hypothesis which has been advanced may, with its corollaries, be summarized as follows: Hughlings Jackson's view that two components, "tonic and clonic," were concerned in the perfect co-ordination of movement has been given experimental verification by the researches of Sherrington. We now speak of tonic and phasic elements in co-ordination. The former, the basis of which is muscle

tone, is concerned with the maintenance and regulation of posture, and between the two elements there is, in Jackson's words, "a perfect co-operation of antagonism." The essential result of a cerebellar lesion is the impairment or abolition of tone, and therefore of all tonic or postural reactions. It is strictly accurate to speak of this as atonia, and in this sense alone we have employed the term. Therefore, in saying that atonia is the essential cerebellar defect-symptom, we express the loss of one of the two essential elements in motor co-ordination. The diverse and conflicting analyses of the cerebellar symptom-complex to be found in the literature describe a large number of so-called elementary defect-symptoms. However, as soon as we adopt the physiological conception of muscle tone and abandon the redundant and empirical nomenclature now employed in this connection, we find that the symptoms which all observers have agreed in describing are nothing but various manifestations of loss of postural tone. In this way we have no difficulty in harmonizing the facts of observation as recorded by different authorities with the hypothesis now put forward.

In this connection it may not be without interest to consider whether what Jackson called a "corresponding opposite" to the combination of loss of postural tone with retention of voluntary power ever occurs in man under the conditions of disease; namely, a state of affairs in which with loss of voluntary power postural tone is preserved. The present writer has suggested elsewhere [18] that such a dissociation of function does occur in spastic hemiplegia where the paralysed limbs show involuntary "associated movements" when the normal limbs are voluntarily put into action. The postural reactions which normally accompany any movement, especially if this be forceful or widespread, are bilateral. In their papers Magnus and de Kleijn have given us many examples of widespread postural reaction, and Sherrington has described the postural accompaniment of the scratch reflex. In hemiplegia the loss of voluntary control over the limbs of the paralysed side cannot result in the suppression of such reactions on that side, but in their "release." Under these conditions we may expect them to develop in an exaggerated form, but lacking in that refined character which they normally possess. It is reasonable to suppose that the associated movements of the paralysed limbs in hemiplegia are manifestations of this nature. If this be so, we have a very familiar "corresponding opposite" to cerebellar ataxy. In both instances the falling out of one of the component elements in the process of co-ordination results in gross impairment of the factors remaining,

whatever be the physiological rank of the lost factor. Thus voluntary movement is disordered from loss of postural tone and postural reactions suffer when voluntary control is withdrawn.

Chapter IV.—Some Theoretical Conclusions.

If we accept this hypothesis as the most satisfactory generalization of the facts of observation, we can readily see how it is that the older analyses have failed to provide the basis for any theory of cerebellar functions. As we have indicated, the most concise analysis is that of Luciani's, but at the time it was formulated so little was known of the nervous processes underlying the maintenance and regulation of muscle tone that it was not possible for him to achieve a final solution of this fascinating and difficult problem. We may now pass to consider very briefly whether the point of view we have adopted in the present paper provides a more promising vantage ground for an attack upon the physiological problems presented by the cerebellum. It would be easy to conclude at once that the cerebellum is the organ for the regulation and integration of all the elements concerned in the maintenance and regulation of posture. Unfortunately, the matter is by no means so simple, and such a view does not take into account some recently recorded observations of Magnus. Our first difficulty is to explain how a defect lesion of the cerebellum produces loss of postural tone. When we compare the results of ablation of this organ in the intact and in the decerebrate animals a striking difference appears. Whereas all observers are agreed that an acute and purely cerebellar lesion in man, or the ablation of the organ in an animal with an otherwise intact nervous system, produce the characteristic cerebellar ataxy, yet, as Sherrington was the first to point out many years ago [10], in the decerebrate animal the rigidity is not abolished by subsequent removal of the cerebellum, nor conversely does ablation of the cerebellum prevent the development of rigidity upon subsequent decerebration. Further, Magnus [6] [7] [8] finds that not only does rigidity persist after ablation of the cerebellum, but all the labyrinthine reflexes which normally play so fundamental a part in the co-ordination and regulation of muscle tone are intact. In other words, the reflex paths concerned in the maintenance and regulation of tone do not pass through the cerebellum. Magnus concludes that we cannot regard the cerebellum simply as the central nervous organ of the labyrinth, and so far as the decerebrate animal is concerned it seems as though we cannot even

regard the cerebellum as essential to the maintenance and regulation of posture. *In short, in the intact animal, the cerebellum appears essential to the maintenance and regulation of postural tone, but in the animal deprived of its cerebral hemispheres it is not essential to either process, as far as either is carried out under these conditions.* The whole question of tone, and the various ways in which it may be affected by different lesions of the nervous system, as we have discussed them, may be simply expressed in the following diagrams. Here the lesion is indicated by a heavy line and the resulting state of postural tone is tabulated in the legend.

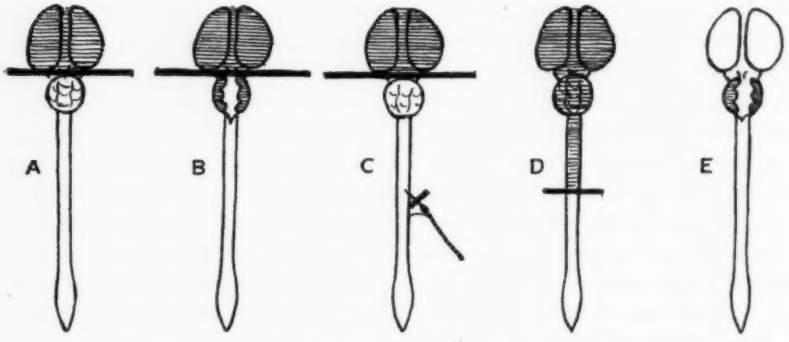

FIG. 1.—A, Decerebrate animal with intact postural tone; B, decerebrate animal with ablation of cerebellum and intact postural tone; C, decerebrate animal with posterior root section and loss of postural tone in "de-afferented" muscles; D, spinal animal with loss of postural tone; E, ablation of cerebellum, central nervous system otherwise intact; loss of postural tone. Heavy lines indicate the situation of the lesion. The portions of the nervous system out of action are shaded. Ablation of the cerebellum, or gross lesions in man, are indicated by shading of cut cerebellar peduncles.

The explanation of this apparent anomaly is probably to be found in certain limitations inherent in these experimental observations. The decerebrate animal is a reflex machine devoid of all volition. Its reflex reactions, although perfectly co-ordinated, consist in relatively simple mass movements, for which the reflex centres situated in the brain-stem are adequate. The voluntary motor activities of the intact animal, and still more those of man, display innumerable variations and elaborations of these simple movements. From their studies of the responses of the anthropoid motor cortex, Leyton and Sherrington [4] draw the inference that it is the function of this region of the cerebral cortex so to influence the lower level reflex mechanisms which are directly con-

cerned in the co-ordination of movement that the skilled, isolated movements of the individual are produced. The cortex by its activity analyses the large and simple reflex movement-complexes into their fractional elements, and synthesizes these into combinations and sequences of almost infinite variety, which make up the normal purposive movements of the individual. We have seen that there are postural and phasic factors concerned in these processes. May we not suggest that the cerebellum is the organ through which the cerebral motor cortex influences postural activities and regulates posture in the interests of co-ordinated purposive movement? It may be the subordinate mechanism employed by the cerebral cortex in the refined correlation of posture with movement.

It has been the custom to think of the cerebellum as a physiological as well as an anatomical entity, though like any other anatomical subdivision of the brain it can have no physiological meaning apart from the rest of the nervous system. That we should continue so to regard it is probably a potent factor in our inability to form any convincing theory of its functions, and a relic of the period when the localization of function in the brain was envisaged with an almost geographical precision. Once we have assimilated the conception of postural and phasic components of movement, and have come to regard the cerebral motor cortex not as itself directly concerned in the processes of co-ordination, but as playing upon the lower level mechanisms which are engaged in this function, and as integrating the postural with the phasic elements of motor activity, it becomes somewhat easier to see why the results of cerebellar lesions should be so different in the intact and the decerebrate animals. In all probability, it is in the light of its functional relationships with the cerebral cortex that we shall ultimately gain an insight into the functions of the cerebellum. Certainly, the parallel development of cerebral cortex and neocerebellum, as revealed by comparative anatomy, points the way to such a conclusion.

REFERENCES.

[1] André-Thomas. "Cerebellar Functions," Journ. Nerv. Dis. Publishing Company, New York, 1912.

[2] Babinski, J., et Tournay, A. "Les symptômes des maladies du cervelet et leur signification," *Trans. XVIIth Internat. Cong. Med.*, London, Section XI, Neuropathology, **1**, 1.

[3] Holmes, Gordon. "The Symptoms of Acute Cerebellar Injuries due to Gunshot Injuries," *Brain*, 1918, **40**, 461.

[4] Luciani, L. "Human Physiology," London, 1915, **3**, 419.

[5] LEYTON, A. S. F., and SHERRINGTON, C. S. "Observations on the Excitable Cortex of the Chimpanzee, Orang-utan, and Gorilla," *Quart. Journ. Exper. Physiol.*, 1917, **11**, 135.

[6] MAGNUS, R. "Welche Teile des Zentralnervensystems müssen für das Zustandekommen der tonischen Hals- und Labyrinthreflexe auf die Körpermuskulatur vorhanden sein?" *Arch. f. d. ges. Physiol.*, 1914, **159**, 224.

[7] *Idem.* "Beiträge zum Problem der Körperstellung: I. Mitteil. Stellreflexe beim Zwischenhirn- und Mittelhirn-Kaninchen," *Arch. f. d. ges. Physiol.*, 1916, **163**, 405.

[8] MAGNUS, R., und DE KLEIJN, A. "Ueber die Unabhängigkeit der Labyrinthreflexe vom Kleinhirn und über die Lage der Zentren für die Labyrinthreflexe in Hirnstamm," *Arch. f. d. ges. Physiol.*, 1920, **178**, 124.

[9] SHERRINGTON, C. S. "Experiments in Examination of the Peripheral Distribution of the Fibres of the Posterior Roots of some Spinal Nerves" (Croonian Lect., R.S., 1897), *Phil. Trans. Roy. Soc.*, 1898, **158**, B, 45.

[10] *Idem.* "Decerebrate Rigidity, and Reflex Co-ordination of Movements," *Journ. Physiol.*, 1898, **22**, 319.

[11] *Idem.* "On Plastic Tonus and Proprioceptive Reflexes," *Quart. Journ. Exper. Physiol.*, 1909, **2**, 109.

[12] *Idem.* "Reciprocal Innervation of Antagonistic Muscles; Fourteenth Note—On Double Reciprocal Innervation," *Proc. Roy. Soc.*, 1909, **81**, B, 249.

[13] *Idem.* "Flexion Reflex of the Limb, Crossed Extension Reflex, and Reflex Stepping and Standing," *Journ. Physiol.*, 1910, **40**, 28.

[14] *Idem.* "Reflex Inhibition as a Factor in the Co-ordination of Movements and Postures," *Quart. Journ. Exper. Physiol.*, 1913, **6**, 251.

[15] *Idem.* "Postural Activity of Muscle and Nerve," *Brain*, 1915, **38**, 191.

[16] THIELE, F. H. "On the Efferent Relationship of the Optic Thalamus and Deiters' Nucleus to the Spinal Cord, &c.," *Journ. Physiol.*, 1905, **32**, 358.

[17] VIETS, H. "Relation of the Knee-jerk and Patellar Clonus to Muscle Tonus," *Brain*, 1920, **43**, 269.

[18] WALSHE, F. M. R. "On the Genesis and Physiological Significance of Spasticity and other Disorders of Motor Innervation, with a Consideration of the Functional Relationships of the Pyramidal System," *Brain*, 1919, **42**, 1.

[19] *Idem.* "The Cerebellar Symptom-complex and its Significance," *Medical Science*, 1921, **5**, 134.

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Persistent Pain in Lesions of the Peripheral and Central Nervous System.

By Wilfred Harris.

Persistent pain, which may be defined as acute bodily discomfort, varying in degree from a sensation of soreness or aching to one of intense and intolerable agony of torment, may be due to a variety of diseases affecting primarily other structures and tissues than the nervous system. Thus, carcinoma naturally occurs to us, or other malignant growth, as a probable cause of any deep-seated pain, persistent in character and lasting for weeks and months, more especially if accompanied by progressive emaciation.

The appreciation of any sensation of pain due to visceral or bone disease necessarily involves the agency of the nervous system in the transference of the sensory impulses from periphery to thalamus and cerebral cortex; but I am limiting my remarks as far as possible to diseases involving the nervous system itself, and shall omit discussion of the referred pains of visceral disease, such as dental neuralgias, errors of refraction, angina, the colics, sinus suppuration, abscesses, acute inflammations and the like.

For the sake of convenience I have in the following classification arranged the causes of persistent pain in diseases of the nervous system in five classes, progressing from periphery to centre, or from nerve-ending to cerebral cortex.

Persistent Pain in Lesions of the Peripheral and Central Nervous System.

(1) *Peripheral due to inclusion of nerve-endings in scar.*

(*a*) e.g., Neuro-fibrositis, traumatic or rheumatic; adiposis dolorosa.

(*b*) Due to septic or to rheumatic terminal trigeminal neuritis:—

(i) Chronic paroxysmal trigeminal neuralgia (Fothergill's disease, tic douloureux).

(ii) Chronic neuralgia of upper or lower jaw.

(*c*) Geniculate neuralgia.

(*d*) Glossopharyngeal neuralgia.

(2) *Disease involving nerve trunks.*

(*a*) Supraorbital neuralgia.

(*b*) Multiple neuritis.

(*c*) Brachial or sciatic perineuritis.

(*d*) Tumours or gummatous neuritis:—

(i) Trigeminal.

(ii) Spinal, e.g., neurofibromatosis.

(*e*) Causalgia.

(*f*) Cervical or first rib pressure.

(3) *Lesions of posterior root ganglia or posterior roots.*

(*a*) Post-herpetic neuralgia:—

(i) Trigeminal.

(ii) Spinal.

(*b*) Tabetic neuralgia.

(*c*) Other root scleroses.

(4) *Central sclerosis of fillet or thalamus.*

Intra-medullary growths.

Syringomyelia and syringobulbia.

(5) *Psychalgias.*

Neurofibrositis.—The pains of chronic or acute fibrositis of the lumbar or dorsal region, often known as lumbago and muscular rheumatism, are doubtless only too well known to many of us, though common as the affection is, its pathology is largely a matter of conjecture. Violent or, and especially, sudden muscular action is the exciting cause in a considerable number of cases: the sudden onset of intense pain immediately after a heavy muscular strain, such as a slip or fall when carrying a heavy weight, the pain relieved by

lying down or sitting in a deep chair and aggravated immediately on movement, suggest rupture of muscle or tendinous fibres involving sensory nerve filaments, as the cause. In the majority the pain disappears after a few days or weeks, while occasionally the disorder persists for years.

Recently I was consulted by a farmer, a powerfully built muscular man, who some ten years ago slipped and fell when carrying a sack of wheat weighing about 260 lb. He was immediately seized with acute pain across the lumbar region, and more especially in the neighbourhood of the right posterior iliac spine. He has been subject to this pain ever since off and on, worse on movement or during heavy lifting, and now and again he is subject to exacerbations so severe he can scarcely move or dress himself. When I saw him during such an attack, his spine was flexed laterally towards the right, and there was a small circumscribed area of great tenderness on deep pressure over the right posterior iliac spine.

Careful marking of the site of chief tenderness, followed by injection of a few minims of 90 per cent. alcohol deep into the tender area, completely relieved his pain within fifteen or twenty seconds, so that he was able to stand up straight, the lateral flexion of the spine having now disappeared, and he walked away in comfort.

Acupuncture has long been recommended since Sydney Ringer's time for similar conditions, but if one or more definite areas of tenderness on deep pressure are discovered, I consider that alcohol puncture is far more valuable.

Traumatic neuro-fibrositis may involve larger nerve trunks, not nerve filaments only, and sciatic perineuritis is a fairly common early sequel to fibrositis of the lumbar region, whether this is of rheumatic origin or due to a fall or other injury. Here again the pain may continue for months or even years. Sciatic perineuritis as a sequel of muscle strain or fall is particularly amenable to treatment by massive saline injections into the nerve just below the notch, the probable site of the nerve sheath damage. Alcohol injection of the sciatic must, of course, never be attempted, owing to the certain result of paralysis of the foot which would ensue, but saline injection of 50 or 60 c.c. preceded by 2 c.c. of 2 per cent. novocain into the nerve often cure the sciatica immediately. This treatment, though often valuable, is less certainly successful in the rheumatic sciaticas, possibly owing to the large area of the nerve which may be the seat of trouble. In both the rheumatic and the traumatic sciaticas there

are frequently spots of deep tenderness with aching pain either in the neighbourhood of the trochanter or between the notch and iliac crest, which may keep up the discomfort and pain after the actual sciatica has been relieved. These are probably local areas of neurofibrositis in the glutei or erector spinæ, and may be mostly successfully injected deeply with alcohol, after the nerve itself has been treated with the saline injections. Probably the saline injections relieve by breaking apart laterally adhesions of the nerve sheath, as the sciatic nerve is a loosely built nerve and readily takes the fluid, swelling up like an egg at the site of injection (as may be seen by injecting the nerve after exposing it on the post-mortem table).

Paralysis of the sciatic I have never seen as a result of violent muscular action, but in the region of the brachial plexus local paralysis from involvement of the posterior thoracic, long thoracic, or circumflex nerves may occur. As with sciatica, so *brachial perineuritis* may occur from muscular overstrain, or rheumatism and other toxic causes, such as pyorrhœa. Usually the posterior cord and musculo-spiral nerve suffer most. As in the lumbar region, so also in the neighbourhood of the scapula and sometimes in the forearm, areas of deep tenderness may be the sole cause of chronic wearing pain, or may accompany a more extensive brachial neuritis. Alcohol puncture of such tender spots often has most brilliant results, chronic pain that may have persisted for many months or years being relieved instantaneously. The spots must be carefully located and marked, and a fine needle, hypodermic size, is usually long enough, plunged vertically into the spot down to the level of the rib or scapula. Care must of course be taken that the needle does not pass between the ribs and pierce the pleura.

Bruising of the scalp by an injury to the vertex from a blow against the lintel of a door, or fall of a heavy weight, is sometimes followed by persistent headache, varying in intensity, or periodic, and associated with a local area of tenderness on pressure. These cases also may be relieved by the same treatment. This form of headache must be distinguished from migrainous neuralgia, in which the periodic headache is unilateral and often associated with intense pain and tenderness on pressure on one temple. Alcohol injection in these cases is quite useless, as is only to be expected, the origin of the pain being probably central and not peripheral.

Chronic paroxysmal trigeminal neuralgia is certainly of peripheral origin, and is probably due to septic neuritis of nerve filaments in the maxilla or mandible. Scarcely ever does this disease affect the

upper division and supraorbital alone, but it may be involved along with the second division.

John Fothergill's original description nearly one hundred and fifty years ago of the painful attacks is scarcely to be improved on to-day, though his view of its pathology, thinking that it must be cancerous, because of the long-standing pain, is now discarded. Many causes appear to contribute to its appearance. Heredity of the disease I have met with in about 1 per cent. of my cases. Strong emotion may precipitate an attack. Injury and blows on the jaw are a not unfrequent cause, as may also be exposure to severe chill, as in motoring or driving; a septic antrum has preceded typical tic douloureux of the second division too often to be a coincidence. I find the disease twice as frequent in women as in men, a point also noted by Fothergill, though he saw only sixteen cases in his practice (or business, as he calls it). A curious point my statistics show is that the disease is twice as frequent on the right side as the left, and commoner in the upper than the lower jaw, though both are often affected. The larger incidence on the right side may be due to better use of the toothbrush on the left side of the mouth, as would be natural in right-handed people, according to Mr. Warwick James. Bilateral trigeminal neuralgia is comparatively rare. I have met with it perhaps thirty times only among several hundreds of cases. The disease rarely disappears spontaneously when established (I have known this occur only once, in the father of a sufferer from this disease, who was my patient). Her father suffered for twenty-five years until he was aged 90, but was free for the last fifteen years of his life, living to 105. In the early stages, however, long periods of remission may occur; I have known thirty years elapse between the first and second attacks, and intervals of one to two years are common. Sometimes a certain periodicity is seen, as for a few weeks to three months every autumn or winter. As the years pass usually the intervals of freedom get shorter. In a few, there is practically no remission, agonizing pain inevitably following every movement of the face, as in eating or in washing. One of my patients, a lady, had been unable to wash the right side of her face for twenty years. Often the lightest touch of a hair or draught of air will start a paroxysm, though at other times nothing may provoke the pain. During the painful bouts the prick of a pin usually seems more acute on the affected side, though this is a temporary hyperæsthesia only, and is not present between the attacks.

Fothergill, 150 years ago, thought hemlock pushed to toxic doses was a cure for this complaint, but our experience nowadays is that drugs comparatively rarely are of service, and that real relief is obtained only by a solution of continuity of the offending nerve trunk. Practically the choice, in severe cases of pain, is between gasserectomy or division of its sensory root, both severe operations, on the one hand, and alcohol injection of the nerve trunks at their deep foramina, or injection of the ganglion itself. For bilateral cases of this neuralgia the gasserectomy operation is not permissible, owing to the jaw drop that would ensue, though bilateral destruction of the ganglion by injection may be done, as the motor root then recovers, though the sensory ganglion cells are permanently destroyed. A curious point I have often noticed when injecting the foramen ovale and ganglion is that the ophthalmic or inner portion of the ganglion goes totally anæsthetic before the second division, and it may be extremely difficult at times to get total anæsthesia of the cheek when the forehead and eyeball remain perfectly anæsthetic. I have no satisfactory reason to account for this.

Another puzzle that occasionally, though fortunately rarely arises, is the partial recurrence of sensation on the chin and remainder of the third division, when the first and second divisions remain totally anæsthetic. With this reappearance of sensibility, pain may return, and that this is not necessarily the result of the faulty injection is proved by the fact that gasserectomy and division of the sensory root behind the ganglion may not alter the conditions. One is tempted then to call the persistent pain a psychalgia, but of this I am not convinced, as there is no doubt true sensation on the lower lip and chin. Gasserectomies, of course, like other operations, may sometimes fail through being incomplete; I have seen several such cases where pain has returned after about ten years, with reappearance of cutaneous sensibility, yet I do not think that is the explanation in the few cases above referred to, though no such case that I am aware of has come to autopsy.

Another form of *persistent trigeminal pain* that I have seen many instances of is in my experience peculiar to young women. It is continuous, not paroxysmal, though it may vary in severity, and it affects either the upper or the lower jaw. It is not provoked by eating, laughing, washing or other movements of the face as is true tic douloureux. It is more difficult to relieve by alcohol injection than spasmodic tic douloureux, inasmuch as total anæsthesia is

necessary to abolish the pain, and with commencing regeneration of the nerve the pain recurs. In tic douloureux, however, in a large majority a medium anæsthesia from injection may be sufficient to abolish the pain, in some cases for many years; in one of my early cases there has been no recurrence 12½ years after injection of the foramen-ovale, though only light anæsthesia now remains.

This lady had previously suffered from typical tic for twenty years, section of the inferior dental nerve having given only two years' relief. In her case the neuralgia commenced at 17, and I have seen it commence at 17 in two other cases and once at 16, many in the twenties, but most commonly about the age of 50. On the other hand the type of persistent neuralgia of upper or lower jaw previously described I have met with only in women of 15 to 35. Its cause I am very uncertain about, unless it is a chronic osteitis of the jaw. Its limitation to women I do not understand, as the sufferers I have met with have not been notably of the neurotic type. Moreover, the true tic douloureux is undoubtedly much more frequent in women than in men, about two to one. These cases are more difficult to treat than true trigeminal neuralgia, as nothing short of total nerve destruction cures the pain.

Supra-orbital neuralgia.—Persistently recurrent supra-orbital neuralgia is met with in both sexes, though oftener in women. I assume that migrainous and true trigeminal neuralgia, and the peripheral causes such as errors of refraction, frontal sinusitis, frontal herpes, antral abscess and dental neuralgias have been excluded. Sometimes paroxysmal supra-orbital neuralgia occurs daily, coming on about the same time, perhaps 10 or 11 a.m., and lasting until 5 p.m. I have seen this type follow influenza several times. Usually the pain is limited to the supra-orbital nerve, but it may involve the whole of the ophthalmic branch. In a recent case, a woman of 47, pain began four years ago, lasting for eight hours daily from January to June. Next year and the year after a similar repetition occurred. A year ago she had a more severe attack and since then has had pain daily from 12 noon to 5 p.m., never a day free. The pain is situated in the forehead and left side of head as far back as the coronal suture, and extends along the left side of the ridge of the nose. Unlike trigeminal neuralgia the pain is not evoked by talking, eating, or other movements of the face, or by rubbing. Supra-orbital injection gave little or no relief, though deep anæsthesia was produced. Injection of the Gasserian ganglion was then done, the anæsthesia of the first division being total

for an hour or more, but subsequently wore off partially. The neuralgia disappeared completely for several days, and then reappeared in a much attenuated form. Very probably total destruction of the ganglion would have produced a complete cure of the neuralgia.

Perhaps the majority of paroxysmal supra-orbital neuralgias are migrainous in type, for which alcohol injection is of little or no use. In some subjects this migrainous periodic neuralgia is limited to the temple.

Only comparatively rarely does true trigeminal neuralgia invade the first division of the fifth nerve, that is to say a paroxysmal neuralgia, sudden and intense, and brought on by movements of the face, rubbing of the skin, or even a draught. In only one instance have I seen it remain limited to the first division of the fifth for years without either the second or third division becoming involved, and even in this case rubbing the chin would start the pain in the forehead. Practically always the neuralgia also involves the second or even all three divisions when the supra-orbital distribution is involved, though it is to be remembered that the pain in the second division of the fifth frequently, indeed generally, extends above the eyebrow and in front of the temple.

In all cases of persistent or recurrent pain, anæsthesia must be looked for. If it is present in trigeminal cases then either syphilitic neuritis, gumma or tumour is the cause.

Geniculate neuralgia, or neuralgia affecting the distribution of the sensory fibres contained in the seventh nerve, has been fully described by Ramsay Hunt and others, though some deny the association of the seventh nerve with such neuralgias. Transient pain around and behind the ear, lasting for two or three days, is a commonplace in association with the onset of facial palsy, and often precedes the motor paresis. Much rarer are instances of true herpes zoster affecting the auricle in association with facial palsy. The distribution of the herpetic rash is usually on the concha and antihelix, though it may be found behind the ear where the pinna joins the scalp, and also along the posterior wall of the external auditory meatus. Such a distribution of herpes I saw once in a medical man, without facial palsy, who for many weeks afterwards had excruciating paroxysmal neuralgia affecting the ear, front of the ear, and back of the lower jaw and neck. Pierce Clark and Taylor, of America, describe a case of chronic tic douloureux affecting this area which was cured by operation by trephining and dividing the pars intermedia Wrisbergi, or sensory root of the seventh nerve, intracranially.

Glossopharyngeal neuralgia.—A rare form of chronic paroxysmal neuralgia or tic douloureux may affect the glossopharyngeal nerve. In its paroxysmal suddenness of onset, and in the severity of pain, glossopharyngeal tic is identical with trigeminal tic, for which it may easily be mistaken. Distinguishing it, however, from the latter, the pain in glossopharyngeal tic starts in the throat, in the region of the tonsil and anterior pillar of the fauces. The pain radiates to the ear, and especially just in front of the ear, along the back of the mandible, and into the upper part of the neck. I have met with two only of these cases, both of which had lasted over ten years, one a man aged 40, the other an old lady of 87. As in trigeminal tic, the first onset of the pain may be furious and sudden, in the man's case the pain striking him in the throat as he opened his mouth to eat a sandwich. When I met with these cases, now ten years ago, I knew of no reference to glossopharyngeal neuralgia, and in case the pain might be an unusual form of trigeminal neuralgia, I injected the third division at the foramen ovale in each case, quite successfully as regards destroying this branch, but without influencing the recurrent neuralgia. I have since seen somewhat similar spasmodic pain, with intense hyperæsthesia of the side of the neck, in a case of recurrent epithelioma in the tonsil, following seven years after extirpation of a vocal cord for malignant growth. It was this case which convinced me of the identity of the glossopharyngeal tic in the two previous cases referred to, and I have lately seen a description by Sicard in France of three cases met with during the war of glossopharyngeal neuralgia, which were cured by surgical help by division of the nerve in the neck.

Diseases affecting Nerve Trunks.

Chronic pain in the distribution of the trigeminal nerve may be due to tumours or gummata involving the sensory root of the fifth nerve within the skull or one or more of its branches externally. Tumours in the ponto-cerebellar angle may irritate the sensory root of the fifth and simulate trigeminal neuralgia for years. The pain however, though variable, is less spasmodic and not brought on by light touches or movements of the face. The main point in distinguishing lesions of the trunk or main branches from trigeminal neuralgia is the appearance of diminished sensibility in the affected area. Motor palsy of the muscles of mastication also may be present. When these signs of gross damage to the fifth nerve are found, trigeminal neuralgia may confidently be excluded unless neurectomy or alcohol injection has

previously been done. Peculiarly distressing cases to deal with are the naso-pharyngeal growths, causing persistent pain and increasing anæsthesia of either the second or third divisions of the fifth. When the growth invades the zygomatic fossa and involves the third division, deafness of that ear is usually produced by involvement of the Eustachian tube. When the growth is more central and invades the spheno-maxillary fossa, besides involving the second division of the fifth, there is produced presently diplopia and later proptosis and fixation of the eyeball through extension into the back of the orbit. Beyond anodyne remedies I know nothing that relieves these cases. Operation seems useless; the growth is never circumscribed and always recurs. In malignant growths of the maxilla pain may be exceedingly troublesome, even after excision, due to recurrent growth. This pain may be completely relieved by a successful alcohol injection of the Gasserian ganglion. Similarly the agonizing pain due to carcinoma of the side of the tongue and lower jaw may be completely arrested by alcohol injection of the third division of the fifth at the foramen ovale.

Persistent trigeminal pain for weeks and months, non-spasmodic, and accompanied by diminished sensibility of that side of the face, but without the pressure signs above referred to of Eustachian deafness, proptosis, and diplopias, should suggest gumma as a probable cause. Even if the Wassermann reaction be negative, it is well always to give anti-syphilitic treatment a trial. I have met with several such cases of syphilitic trigeminal neuritis which cleared up completely under treatment by biniodide of mercury or salvarsan injections.

Rib Pressure on Brachial Plexus.

Chronic pain in the arm and neck, running down to the inside of the hand, in women between 20 and 30, will usually suggest cervical rib as a cause through pressure on the first dorsal nerve, as it rises to join the inner cord. Wasting of the musculature in the hand and diminished sensibility along the inner border of the forearm to the wrist render the diagnosis more certain. If a skiagram demonstrates a cervical rib, the position is clear. Many cases of rib pressure due to the first rib alone, in the absence of a cervical rib, have been recorded; I have had two such cases successfully relieved by operation within the last six months. In one of them the first rib caused a prominent hard swelling above the clavicle which it was difficult to believe was not a

cervical rib when the skiagram proved the absence of the latter. The diagnosis is therefore more complicated and difficult now that we must realize that with symptoms suggestive of cervical rib the skiagram may be normal.

Causalgia.

During the war the frequency of cases of persistent agonizing pain due to injuries of nerves, often slight, was most remarkable. The large majority of these cases involved the median or internal popliteal nerves; though I have seen it also in the distribution of the ulnar, long saphenous, external cutaneous of the thigh, and radial nerves.

In some cases the nerve injury was so slight that no demonstrable anæsthesia was present—only an intense hyperæsthesia, varying at times, and liable to be aroused into an intense spasm of pain by a sudden emotion, noise, or vibration or light touch, thereby reminding us somewhat of the onset of the paroxysms in trigeminal neuralgia. The pain is described as of a burning heat—hence the name "causalgia" (*καιω*, I burn), and bursting sensations in the fingers were common. The pain in many cases lasted for weeks and months, and in some it was necessary to produce nerve-blocking, in order to arrest the pain, by alcohol injection of the trunk of the nerve above the injury. Common enough during the war, I have seen only one such case in civil life where a man tore his thenar eminence in some cog wheels two years previously. The pain was limited to the ball of the thumb, and immediate and complete relief was obtained by injecting the median nerve with alcohol just above the wrist. The nerve can be easily found here, as it lies between the tendons of the flexor carpi radialis and palmaris longus, and pricked with a hypodermic needle and injected. No other operation is necessary for this.

Painful stump was another very common sequel of amputation in the war.

Brachial and Sciatic Perineuritis.

Sciatica I have already referred to, especially in association with lumbo-sacral fibrositis, either of a rheumatic, septic, or traumatic origin.

A very chronic and obstinate type I would like to refer to here in which the most noticeable sign on examination is the scoliosis, a lateral flexion of the spine away from the painful side, causing the hip on the affected side to stick out. The lower cervical spine may be as much as 5 in. to one side of the vertical median line when standing. This scoliosis disappears on sitting or lying. It may be a result of a local

lumbar fibrositis in the erector spinæ in the region of the fourth and fifth lumbar transverse processes. Occasionally a definite area of deep tenderness can be localized and it is sometimes possible to procure an immediate and dramatic relief by alcohol puncture of the spot. Short of that treatment these cases are very chronic and resistant to ordinary treatment, and three months' rest in bed may be necessary.

Brachial neuritis is seldom so chronic as some of the sciaticas, though the acute form is probably more painful, due I think to the structure of the cords of the brachial plexus being denser and firmer than the sciatic and thus admitting of less swelling in inflammation of the sheath. This point is easily demonstrated in the dead body by cutting down on the sciatic and brachial nerve trunks and injecting fluid through a hypodermic needle. The sciatic takes it easily and swells up in an egg shape, but it is difficult to force more than a few drops into the main brachial trunks.

Chronic pain about the scapula and arm may be due to cervical rib, or to neurofibrositis, as already stated. A common cause of pain referred from the shoulder down the arm, perhaps to the elbow, its maximum usually about the insertion of the deltoid, is adhesions in the shoulder-joint, and resulting from some slight injury such as strap hanging, being jerked by getting on or off a bus or tram, or by a slight fall.

These cases are frequently diagnosed as neuritis. The pain starts usually a day or two after the injury and is due to a synovitis, the subsequent adhesions causing pain with every movement or pressure on the joint. Wrenching under gas, with subsequent passive movements and massage, usually effect a cure, but some cases are too painful to tolerate the after treatment and partial fixation is permanent. To be distinguished from this condition is the fixation of shoulder from adhesions subsequent to immobilization during the acute stage of brachial neuritis. The history of severe pains and pins and needles down to the fingers generally makes this point clear, but the treatment by massage, passive movements, or even wrenching under gas is the same.

Disease of Posterior Spinal Roots or Root Ganglion.

Post-herpetic neuralgia is one of the most inveterate and difficult neuralgias we have to treat. Due in part to an inflammation lesion in the root ganglion, in some cases to a neuritis of the nerve trunk, and in others even to inflammatory changes in the grey matter of the posterior

horn in the spinal cord, the pain is constant and wearing, causing great depression. Rare under the age of 50, chronic neuralgia following an attack of shingles is increasingly common in older people. In some the pain may persist for months and then gradually disappear, but in others it may remain for years and has led to suicide. With the pain is usually a severe numbness and constriction, and the area of scarring is usually partially anæsthetic. Beyond local anodynes and mild tonic treatment I hesitate to suggest more active measures. Alcohol injections in spinal cases is, I believe, quite useless, and division of posterior roots after laminectomy is, I believe, not always successful. I should be glad to hear experiences of surgeons on this point.

Herpes of the trigeminal area is mostly limited to the ophthalmic division, often called frontal herpes. A common sequel is numbness, sensation of constriction and paræsthesiæ of formication, not really amounting to pain. Occasionally, however, in old people subsequent neuralgia in the anæsthetic area is very distressing and persistent. For this, alcohol injection of the Gasserian ganglion may give complete relief, and I have done this in three such cases, but it is necessary to produce total and lasting anæsthesia equal to that resulting from a gasserectomy.

Chronic pain in the limbs, usually the lower limbs, may result from posterior root sclerosis, subsequent to meningitic lesions. *Hæmatorachis* or intraspinal hæmorrhage in the lumbo-sacral region may result from heavy muscular effort. The effused blood is apt to clot around the roots of the cauda equina and cause chronic irritation and pain in one or both lower extremities, with wasting and loss of reflexes, and possibly diminished sensibility. Lumbar puncture will clear up the diagnosis, a straw-coloured cerebro-spinal fluid being drawn off even a year or more after the onset. In one such case I had laminectomy performed and a large purple clot 4 in. long surrounding the roots of the cauda equina removed, eighteen months after the injury.

Tabetic pains are too well known to need enlarging on here. Their treatment is perhaps less satisfactory. Mercurial friction improves some, intravenous salvarsan yet others, while for those who do not notably improve under salvarsan only I have seen considerable numbers benefit immensely as regards severity of pain by intraspinal injections of the serum taken from the patient's blood after intravenous salvarsan.

Usually there is a strong reaction about two hours after the injection of 50 to 55 c.c. of the serum, severe pains in the limbs coming on and lasting twelve to twenty hours, followed by more or less complete

relief, which may last for years. Presumably the injection of the serum sets up some congestive state in the posterior nerve roots which acts beneficially on the chronic syphilitic neuritis which is the source of the pain. Root sclerosis may be due to other causes, such as hæmorrhage, toxic degeneration in diabetes, and other causes of neuritis.

Proceeding centrally, we find chronic neuralgic pain resulting from intramedullary lesions affecting the fillet and thalamus. Usually the pain is constant, burning, and pins and needles in character, but occasionally it is paroxysmal and neuralgic. More than ten years ago I showed before the Neurological Section a man of over 60, who had suffered from an attack of thrombosis of one posterior inferior cerebellar artery. As a result of the thrombosis implicating the side of the medulla with the fillet and the descending or spinal root of the fifth nerve, there was analgesia of the left fifth area and of the right half of the body, excluding the face and the forehead. In the analgesic area of the left fifth he complained of constant neuralgic pain, which nothing appeared to relieve. It is perhaps difficult to understand how a simple sclerotic lesion can cause persistent neuralgia in the absence of any irritating focus, and this is the only instance of this posterior inferior cerebellar syndrome which I have seen with permanent neuralgia resulting, though this particular thrombosis does not appear to be very rare, as I have met with twenty or more cases. It has been suggested that the persistent burning pain and paræsthesiæ in lesions of the thalamus or fillet is due to the spontaneous unrestrained activity of these nuclear centres for sensation. Similar pain is met with in some cases of syringomyelia and syringobulbia, pain referred to analgesic areas or analgesia dolorosa. Intramedullary spinal tumours also are liable to cause a burning pain as an early symptom, which may precede for many months any more definite localizing signs.

Psychalgia.

Pain of mental origin is usually distinctive in character, such as the vertical pressure pain, or clavus hystericus of some neurasthenic headaches. A mental neuralgia may usually be distinguished from a true neuralgia of peripheral origin by its distribution not being anatomical in form and overlapping other nerve areas, and especially in crossing the middle line. A parson, aet. 63, for 2½ years has had pain in the right great toe. Of three surgeons who saw him, one operated for exostosis, the second excised the joint, the third told him he had a kink in his colon, but let him off with an abdominal belt and paraffin

internally, and advised him to get a tendon cut. In spite of all treatment (or because of it) his pain is now much worse. On inquiring into his history, I found he was the child of first cousins, and that insanity was very prevalent in his family. As I could see nothing wrong with his foot or leg, I have little doubt that this pain was an instance of psychalgia. Occasionally there may be difficulty in diagnosis, as in a Jewish patient whom I saw for pain on the left side of the cheek and forehead and nose, but crossing the nose as far as the inner canthus of the opposite eye. He was anxious for injection treatment, and as he had had much medicinal treatment without benefit, somewhat against my better judgment I injected his second division. Although dense anæsthesia resulted the pain was not improved, rather worse. I then hesitated between advising suggestion treatment and injection of the ganglion. He would not have the former and again against my better judgment I injected him, this time through the foramen ovale, and producing total and permanent fifth nerve anæsthesia. His eye and cornea fortunately gave no trouble, but he was now even more complaining of the pain, and he went to see another surgeon regarding gasserectomy, who, however, refused to do the operation when he found the fifth nerve area totally anæsthetic. Baulked in this he went to yet another surgeon who operated to remove his ganglion; and the last I heard of him was that he was still complaining violently of pain.

The moral of this tale is that it is unwise to attempt any form of surgical treatment for psychalgias; the pain is apt to get worse, or spread to another area, and once an operation or injection has been performed, it is most difficult for another practitioner to sift the real from the false and make a diagnosis. With the psychalgias may perhaps be included many of the *coccygodynias*, though perhaps in the majority of them there is a history of some local injury at the outset.

In conclusion, I must apologize for the discursive nature of my remarks as I have endeavoured to point to special types and difficulties which the combined talent here to-night will, I hope, throw light upon.

On Persistent Pain.

By Sir WILLIAM THORBURN, K.B.E., C.B., C.M.G., F.R.C.S.

THE treatment of a symptom rests always upon a more uncertain basis than does the treatment of a lesion. More especially is this the case with such a symptom as pain, which is not only purely subjective but the nature and intensity of which is not capable of measurement or even of definite expression.

Hence the estimation of the results of treatment requires considerable caution. On the one hand, such results may be due merely to suggestion, as was probably the case with alleged "cures" of tabetic pains by suspension or by stretching of the sciatic nerves—methods which I take to be now quite obsolete. On the other hand an operation which has certainly removed the pain to which it was directed—such as gasserectomy for trigeminal neuralgia—may be followed by the most disagreeable phenomena of "psychalgic" origin. Thus I have seen removal of the Gasserian ganglion followed by a functional hemianæsthesia, and in two typical cases of rib-pressure removed by operation the patients shortly developed abdominal troubles associated with a well defined movable kidney. As moreover many of the cases operated upon for persistent neuralgia are the victims of alcohol, morphia or other narcotic drugs, the picture is often obscured by such conditions, and again the judgment of operation results may be thereby rendered extremely difficult.

We are, therefore, on the safest ground when we are attacking a quite definite lesion, as in removing a source of pressure. We are less certain of results when dealing with root ganglia, whether spinal or cerebral: and we are on the most doubtful ground when dealing with diseases of obscure pathology such as are many of the neuralgias of the limbs.

With regard to pressure lesions, including those of the dome of the thorax, we are on very safe ground, and in nearly all, if not all, cases the removal of the cause will prove curative. The treatment thus resolves itself simply into the recognition of the cause. It is, however, possible, if not probable, that in cases of long standing there has been produced a local neuritis with cicatricial changes which may lead to permanent symptoms, although pain is seldom prominent among these.

With regard to pain due to external injury, the most typical

examples are those due to end-bulbs after amputation. The relief of such pain is generally immediate if the bulbs be excised, but in some cases little or no benefit will follow and even high resection of the affected nerves will fail to cure. In such cases there is again probably an extensive septic neuritis, as described especially by Corner, and for some of them posterior rhizotomy appears to offer the only hope of relief. A good deal of controversy has arisen as to the prevention of such end-bulbs, but my own opinion, founded upon a considerable experience of amputations dating back to a time when many were not aseptic, is that their essential cause is simply sepsis and that section of the nerves at a high level at the time of amputation is a sufficient prevention even in septic wounds. I have not practised any of the methods of occluding the cut ends of the nerves and I am not aware that their omission has given rise to trouble. Upon this point, however, I should greatly like to hear the experience of those who have worked in British hospitals during the war.

With regard to causalgia again the views of those who have had British war experience would be most welcome to me. So far as I can judge Sicard's method appears to be most popular, although I have no personal experience of it. It has, however, one fairly obvious limitation. When the injury lies near to the trunk the neuritis may well have extended so far that it is impossible to inject the nerve above it and it would then again appear reasonable to fall back upon rhizotomy. Similarly resection and suture may quite fail to cure and may leave rhizotomy as our only hope. (Platt found that of twenty cases treated by resection and suture sixteen were cured and one failed even after repetition of the operation, while of three treated by neurolysis only one was cured.)

Rhizotomy is however by no means always effective in this and other severe forms of neuralgia of the limbs. The analysis of Foerster and of various British surgeons given in the *British Journal of Surgery* yields fifty-nine cases of which twelve died or were not traced, while twenty were cured, and in twenty-seven there was either no relief or such relief was partial only. Of two cases of my own one was little if at all relieved, and it is interesting to note that this was a case of avulsion of the brachial plexus in which there was no open wound or septic infection, and in which it was obvious at the time of operation that the roots were matted together by a cicatrix, and that some cicatricial tissue extended into the spinal cord. Here we have a probable clue to the many failures, viz., an extension of disease beyond the posterior root-ganglia.

Before leaving the question of injuries of nerves, I should like to refer to the often very painful cicatrices of the scalp. Why these should cause much pain—sometimes ensuing long after the original injury—is not obvious, but I am strongly inclined to attribute it to attachment of the mobile scalp to the skull and I have certainly found that freeing of the scalp, with or without interposition of some material such as cargile membrane or aseptic wax between it and the bone, is followed by the relief of pain and of other nervous symptoms often associated with it.

Turning now to the brachial, sciatic and other neuralgias of more obscure pathology, these are of course widely treated by non-operative measures or by such measures as acupuncture, saline injections and "bloodless" stretching. Many of the cases recover as do many under the influence of rest alone, and I do not wish to dispute that all these methods have been followed by good results. But in the cases which come before me they have usually been tried and have failed. Hence perhaps I am strongly prejudiced in favour of a full exposure of the nerve. Such an exposure is free from risk and often reveals a definite lesion such as a source of pressure or a perineuritis with light adhesions. Neurolysis, doubtless associated with some stretching, is at least as likely to be curative as the more obscure attacks of apparently minor surgery, while it certainly provides a more complete and logical line of treatment. In many, but by no means all cases, it has given very good results.

Lastly, I must refer to ganglionic and tabetic pains.

As regards trigeminal neuralgia the various modern methods are so well known that it is unnecessary to do more than briefly refer to them. There are two great classes of operation. Schloesser's method of injection and Hutchinson's method of removing the two lower thirds of the ganglion have the merit that, so far as I know, they never imperil the eye, and in my own experience Hutchinson's operation has always been permanently curative. It would, however, be useless in the rare cases in which the ophthalmic division is involved. On the other hand, Haertel's method of injection, total ablation of the ganglion and resection of the sensory root have, to my knowledge, all caused loss of the eye even when performed by the most competent operators. Haertel's injection is the simplest and the safest as regards life, but appears to me the most uncertain and does not commend itself to one who likes to see what he is doing. Division of the posterior root—especially by Adson's technique—will probably supplant total gas-

serectomy, but so far I have always been satisfied with Hutchinson's method.

Post-herpetic neuralgia I have found surprisingly disappointing, but both my own and the recorded cases of root resection for this condition are too few for safe generalization at this time. Rhizotomy would appear to offer the best hopes of cure, but here again it may well be that changes have extended above the level of the root ganglia.

Tabetic pains are becoming increasingly rare, probably as a result of the better early treatment of syphilis. Foerster and Hey Groves have between them collected seventy cases with seven deaths, fifty cured or greatly improved and thirteen failures. Such results are well worth obtaining, especially as the death-rate is very much lower in the hands of surgeons accustomed to laminectomy, and there is also not a little evidence that spinal drainage, *per se*, is of value in locomotor ataxia. Moreover, several of the failures have been clearly due to a too limited resection of roots. Such resection, to be effective, should probably include at least the fourth, and eighth or ninth thoracic roots in cases of gastric crises and should generally be bilateral. I am quite satisfied that, if it be thoroughly carried out, we have in this operation one of real value which ought to be considered in every case of tabetic pain and which ought in the hands of experienced surgeons to be as safe as an exploratory laparotomy.

DISCUSSION.

Dr. GORDON HOLMES said that it was the teaching of English neurology that most diseases of the central nervous system which did not involve the meninges or extend to the posterior or other sensory roots ran a painless course. That that doctrine was true in the majority of cases there could be no doubt, but he was afraid it had been applied rather too widely. It was generally assumed that spinal diseases could produce pain only when the posterior roots were involved, but as a result of his war experience of gunshot injuries he thought it must be regarded as a possibility that a traumatic injury of the pain-conducting tracts within the spinal cord could produce pain, persisting for several weeks at least. It was not so rare as it was generally assumed to be for intramedullary lesions of the cord to produce pain. Perhaps the most interesting point from the theoretical side concerned the manner in which these pains occurred. It was an old hypothesis that the lesion irritated the pain-conducting fibres, and therefore gave origin to the pain which was peripherally referred or projected. He did not think that many of them were willing to accept that explanation now. He had found in certain gunshot injuries of the spine that not merely painful or uncomfortable sensations were

produced, but also an excessive sensation of what might be called pleasure on the affected parts of the body. The easiest working hypothesis in discussing the nature of the central pain was to assume the view put forward years ago by Long, that the pain-conducting fibres throughout the central nervous system were represented by chains of neurons broken up repeatedly in masses of grey matter, and that the pain was due to some structural or dynamic change in portions of grey matter in which the pain-conducting fibres were normally interrupted.

Dr. S. A. KINNIER WILSON said that if it was the case that the tic movement of a tic douloureux disappeared when the neuralgia was relieved—whether it was a tic or spasm was for the moment immaterial—it seemed as though certain inveterate cases of torticollis might be treatable by the relief of the accompanying occipital neuralgia by operation on the posterior root ganglia. Torticollis was a very interesting subject, and often cases of torticollis were particularly untreatable. He also referred to the description in Dr. Harris's paper of certain patients whose pains came on only at certain times of day (for example, a woman who had the pains from 12 to 5 o'clock each day). These cases presented very interesting problems from a theoretical point of view. The view he himself took was that such cases were essentially psychogenic. He did not believe that any factor except one of that nature could produce the recurrence of pain over the same period of hours every day. He added that he had used ionization successfully in several cases of post-herpetic neuralgia when other forms of treatment had been unsuccessful.

The PRESIDENT said that his own experience of posterior root section had been extremely disappointing, and on very few occasions had it been worth doing; especially was this true of doubtful cases of persistent pain of causalgic character. Every kind of surgical treatment for the relief of these pains had been tried and all had had a measure of success, but on the whole there was more disappointment than otherwise. Unless the nerve could be dealt with at a high level and at an early stage there was little likelihood of any great result from alcohol injections, resection of the nerve, or anything of the kind, and after the pain had persisted for years, then the posterior section itself had very little influence.

Dr. WILFRED HARRIS, in replying to the point raised by Dr. Wilson as to the periodicity of the pain, would not subscribe to the opinion that the periodicity meant psychalgia. Periodic pain was familiar to many of them in other connections. Moreover, the pain in this case had an anatomical distribution, and, contrary to the usual results in psychalgia, injection of the ganglion produced almost complete relief.

CLINICAL MEETING HELD AT THE HOSPITAL FOR PARALYSIS AND EPILEPSY, MAIDA VALE, DECEMBER 8, 1921.

Dr. WILFRED HARRIS, Vice-President, in the Chair.

(1) *Case of Division of Right Sciatic Nerve six years ago; Suture of Nerve six weeks later. Muscles show no wasting, but the Response to Faradism is diminished, and Voluntary Contraction is absent. The Calf Muscles are in spasm.*

By Dr. JAMES COLLIER.

W. H., aged 31, ex-soldier.

History.—Bullet wound severing right sciatic nerve in upper third of thigh July 30, 1915. Operation September 9, 1915, nerve found completely divided, ends sutured.

Discharged from hospital in June, 1916. Patient states there was wasting in right lower leg at this time. No treatment since, but patient always kept on his legs with the aid of a stick. By February, 1917, right leg had regained normal contour and measured practically the same as the left.

Admitted National Hospital, November 29, 1921.

Present condition.—Calf measurements at their thickest girths both 15 in. Muscles of right calf feel firmer than those of left. Constant fibrillary tremor of right calf muscles. Response of these and the fore-tibial muscles to faradism present with a strong current. No voluntary movement of right toes or at ankle, good movement at knee-joint. Sensibility to pin-prick is abolished from the lower third of the thigh downwards.

(2) *Post-encephalitic Lenticular Rigidity.*

By Dr. GRAINGER STEWART.

H. S., aged 9 years.

History.—Eighteen months ago patient had an illness during which he was in a sleepy condition for six weeks; could always be roused; both eyes turned inwards for two to three weeks; seemed unable to see for about a fortnight; had pain in back of head.

After drowsy state passed off, he began to have violent choreic movements of arms and head; legs slightly affected; right side worse than left; head turned to right. Movements lasted six months; as they gradually ceased, right side became weak.

Present condition.—Looks healthy. Face almost expressionless when at rest, quickly lighted up by smile.

Walks with body bent forward, tends to run. Cannot stand still without effort, but tends to fall backwards.

Mental condition: Very bright. Rather sly and precocious; often disobedient; sometimes sulky.

Signs: Pupils equal, react well to light, badly on convergence. External ocular movements full; deviation to left accompanied by nystagmus.

Fundi normal.

Hand-grip and arm movements slightly weaker on right side.

Resistance to passive movements of right upper and lower limbs.

Tendency to Parkinsonian posture.

Knee jerks and ankle jerks present.

Plantars probably flexor, but brisk voluntary extension movements usually occur.

Speech: Often high-pitched intonation—this is not constant and is probably due to habit.

(3) *Gummatous Spinal Meningitis.*

By Dr. A. FEILING.

F. D., female, aged 39, single, was admitted complaining of loss of power in the right shoulder.

Three years ago she had an attack of rheumatism with severe headache and pain in the tip of the right shoulder.

Two months ago pain in the right shoulder again.

Six weeks ago noticed difficulty in raising the arm above the head, and weakness of the arm increased rapidly.

On admission, October 5, 1921, pupils, discs and cranial nerves normal.

Well-marked muscular atrophy and weakness of muscles of the right shoulder, especially deltoid, spinati, biceps, and to a lesser extent of the pectorals, rhomboids and supinator longus muscles. No fibrillary twitching seen. No loss of sensation. Left arm not affected. Tendon reflexes in the right arm not obtained with the exception of a feeble triceps jerk. Only other abnormal signs found were absence of the abdominal reflexes and a double extensor plantar response.

Wassermann reaction in the blood strongly positive. Cerebrospinal fluid, Wassermann strongly positive. Cell count 40 per c.mm. Nonne-Apelt test for globulin positive. Slow but steady improvement is taking place under treatment.

(4) *Disseminated Sclerosis (with Chief Incidence on the Pons).*

By Dr. A. FEILING.

P. L., male, aged 31, complains of weakness of the left side of the face, loss of power in the right arm and right leg.

History and present condition.—August, 1916, fell from a motor lorry; no loss of consciousness. Next day felt "tingling" in the right arm and right leg; within twenty-four hours left facial paresis had appeared; three to four days later marked weakness of right arm and right leg; occasional severe

headaches, no vomiting. Two weeks later deafness in the left ear and left tinnitus. About this time precipitancy of micturition and occasional incontinence of both urine and fæces occurred: also loss of sexual power.

Condition has remained practically the same in last two years: at times the power seems to go from his right arm and leg, with cold feelings in the right side of the body. Occasional headaches; transitory diplopia.

On examination: Pallor of both optic discs. Nystagmus, especially on looking to the right. Diminution of sensation in and paresis of the left side of the face.

Well-marked right hemiplegia with spasticity of arm and leg: tendon reflexes on right side exaggerated; right plantar response extensor, left doubtful, possibly also extensor. All forms of sensation diminished in right arm and right leg.

Sphincters: "Precipitancy of micturition" well marked; no incontinence while in hospital.

Wassermann reaction negative in blood and cerebrospinal fluid (February, 1921).

(5) *Syringomyelia, with Predominantly Unilateral Signs.*

By Dr. A. FEILING.

R. A., male, aged 36, admitted to hospital complaining of loss of sensation in the left hand and arm and weakness of the left arm and left leg.

Two years ago blisters began to form on the left hand.

Eighteen months ago weakness left hand was noticed and the left leg occasionally gave way; loss of sensation in the left hand began at this time.

Constant sweating of the forehead has occurred during the last year. No pain.

On examination: Markedly excessive sweating of both sides of the forehead and on the left side of the face. On the left side the pupil is small, the palpebral fissure narrowed and the eyeball sunken.

Muscular atrophy is seen in the left hand and forearm, in the right shoulder girdle and slightly in the right hand and forearm. Much muscular weakness of left hand and forearm. Trophic lesion of left hand. Both legs are spastic, but especially the left.

Reflexes: Tendon reflexes in the arms are lost, with the exception of a feeble triceps jerk on the right side. Abdominals present on the right and absent on the left side. Knee jerks and ankle jerks exaggerated both sides; ankle clonus on the left side. The plantar response on both sides is extensor. There is prominent kypho-scoliosis.

On the left half of the body sensibility to light touch, heat and cold is diminished; sensibility to pain on the face, trunk and lower limb and abolished on the upper limb. Recognition of changes in passive position and of vibration (C_{1-8}) and appreciation of deep pressure is gravely disturbed in the left upper limb. There is no loss of sensibility on the right half of the body.

(6) *Involuntary Muscular Movements. Torsion Spasm.*

By Dr. A. FEILING.

A. W., aged 13½ years, a Jewish boy, came under observation in July, 1921, for involuntary movements of the head and arms.

He had suffered from an attack of measles as a baby.

No history of any similar complaint nor of any nervous disease on either side of the family.

Symptoms were first noticed when the boy was between 4 and 5 years of age. He would then fall forwards off a chair in which he had been placed. The movements began gradually, first in the neck, and later in the arms, when the peculiar method of grasping an object was noticed. He has been in about the same state since the age of 8. Went to school and has always been mentally alert and bright. Speech normal except for interference caused by the movements.

A healthy boy without signs of disease in heart, lungs, or abdomen. Beyond the involuntary movements the only positive signs are: (1) A lateral nystagmus; (2) inco-ordination of the arms.

No rigidity; no actual paralysis; no loss of sensation; no alteration in the reflexes.

Wassermann reaction in blood is negative.

(7) *Double Ophthalmoplegia Externa (? Congenital) with Generalized Muscular Atrophy.*

By Dr. A. FEILING.

R. M., aged 13 years, male, was brought to the hospital for "weakness of the legs."

Previous history.—Sat up at 12 months, walked at 18 months, talked at 2 years. At age of 2½ years, after an attack of measles, "went off his legs."

Chief complaint is of weakness of the legs, so that he becomes very rapidly tired after walking a short way and has to rest before being able to proceed.

On examination, a small undersized boy. He presents a condition of generalized lack of muscular development with marked weakness of all muscles, especially those of the trunk: thus he was unable to sit up in bed without assistance when he was first in hospital. In addition there is a condition of double partial external ophthalmoplegia, with partial ptosis and defective ocular movements in all directions.

There is slight nystagmus. Vision: right $\frac{6}{9}$, left $\frac{6}{6}$. Fundi normal. No alteration in reflexes; no loss of sensation.

Wassermann reaction negative in blood and cerebrospinal fluid.

(8) *Involuntary Movements, a Sequel of Encephalitis Lethargica.*

By Dr. D. McALPINE. (Dr. WILFRED HARRIS.)

Mrs. M., aged 34. In November, 1920, began to suffer from insomnia; on December 14 she saw double and the diplopia persisted for some days. On December 15 drowsiness developed, which later became marked; she slept day and night. Admitted to Maida Vale Hospital, December 23, under the care of Dr. Wilfred Harris.

Examination showed patient to be very drowsy, but she could be roused and answered questions rationally; speech slow and monotonous; no cranial nerve or ocular palsies: no diplopia; no myoclonic or other movements. During January, 1921, she improved. In March she became more drowsy and involuntary movements were first noticed; these have persisted with varying intensity ever since. She relapsed in June and developed an acute arthritis of the left shoulder-joint as a result of the violence of the movements. There was a further slight relapse in September when a divergent strabismus with loss of power of convergence and defective accommodation were noticed and this condition is still present. At no period has there been any evidence of involvement of cranial nerves or of pyramidal tract; the deep reflexes, sluggish at onset, are now brisk. Plantar responses have always been flexor. The involuntary movements which are now present are rhythmic and uniphasic, occurring about fifteen times to the minute, each spasm lasting about one second. They occur synchronously in the various parts affected. They vary slightly in intensity and regularity, but unless controlled medicinally they are extremely violent and painful and make the patient's life a misery to her.

She can inhibit them to a certain extent, but the next spasm is reinforced. They are increased by emotion and disappear entirely during sleep. Muscular tone in the limbs, in the intervals between the spasms, is not increased.

During an involuntary movement the chin is turned slightly to left; the occiput is retroflexed and approaches the right shoulder. At first, the angle of mouth on the left side was drawn downwards, but now there is usually no such movement. Left arm is internally rotated and pulled slightly backwards, extended at elbow and wrist; forearm pronated and fingers powerfully flexed into the palm. There is shrugging of right shoulder and slight abduction of whole of right arm. The trunk is flexed to the left. The left leg is slightly flexed and sometimes abducted at hip; the knee is powerfully extended; the feet and toes point downwards and the foot is slightly inverted. In the right leg there is slight contraction of hamstrings and dorsiflexion great toe.

Luminal alone of the hypnotics that have been tried seems to control the movements; the dose required is 6 gr. daily. This amount has been given for the last six months and, apart from slight paresis of accommodation, no toxic effects have been observed.

(9) *Involuntary Movements, Sequel of Encephalitis Lethargica.*

By Dr. D. McALPINE.

J. T., aged 52, became ill on December 9, 1919. Pain back of neck. Slept for two weeks; delirious. Afterwards found he could not read properly;

also had difficulty in speaking and weakness in left arm and left leg. In January, 1920, involuntary movements began which have persisted since; left shoulder first affected.

Pupils equal; react normally to light and accommodation. No ocular palsies. Speech slow and slurring. Cranial nerves normal. Slight loss of power left arm. Reflexes and sensation normal in upper and lower limbs. The involuntary movements affect chiefly left face and left arm. They are uniphasic and rhythmic, occurring about sixteen times to the minute. Each spasm lasts about one and a half seconds. They disappear during sleep. They occur at irregular intervals, two following each other often in quick succession; movements of arm sometimes occur without any accompanying movement in face. Peripheral stimulation and emotion increase the amplitude of the movements. In an involuntary movement the head is laterally flexed to left. The chin is depressed, the left angle of mouth is drawn outwards and left cheek sometimes sucked in. The left shoulder is slightly raised, the whole arm being pulled backwards, with the elbow extended, the forearm pronated, and the wrist slightly flexed. The index finger is either flexed or extended, but the middle finger constantly flexed and the thumb abducted, Movements in the arm are often not so complete as those described above. There is occasionally slight contraction of the extensors of left knee. The involuntary movements do not involve the right arm or right leg.

The patient has benefited slightly by the administration of hyoscine; small doses only of luminal have been tried without apparent benefit.

The main features in these two cases are strikingly similar; the nature of the movements seems to indicate a "release" phenomenon; the site of the lesion or lesions is problematical; the absence of rigidity in the interval between the tonic spasms practically excludes a lesion of the corpus striatum. which combined with the fact of absence of cranial nerve involvement seems to point to a lesion in the subthalamic region.

(10) *Case of Cerebrospinal Syphilis.*

By Dr. H. MAUDSLEY. (Dr. FEILING.)

A. W., aged 56, labourer. Married, with two children alive and healthy; wife has had six miscarriages.

He was infected with syphilis thirty years ago. Has always felt well up to onset of present condition. Fairly rapid onset eight months ago of weakness of left hand and forearm. First noticed that he was unable to approximate thumb to his fingers on the left side—the disability gradually progressed with weakness of the hand-grip and wasting of the muscles of the hand and partial loss of power in the forearm and upper arm.

On examination his pupils are found to be equal in size, but slightly irregular in outline; they react very sluggishly to light, but briskly to accommodation. Other cranial nerves are normal.

There is evident flattening of the thenar and hypothenar eminences of his left hand together with some wasting of interossei. He can flex his inner two digits, but is unable fully to flex his thumb or outer two fingers. His power of extension of wrist is fairly good, but he cannot fully extend the distal phalanges. There is some loss of pronation and supination and flexion of left forearm and slight general weakness of the upper arm. There is also a certain amount of wasting in forearm and upper arm. The muscles of the wrist and hand are hypotonic. The power of the right upper limb is good. There is no weakness of his lower limbs.

Upper limb reflexes on both sides are fairly active. Knee-jerks present, left slightly greater than right. Ankle-jerks: left is just present, right absent. Abdominal reflexes are absent left side, just elicited on right side.

Plantars: Double extensor. No clonus and no spasticity of lower limbs.

Sensation: Apart from a slight defect in the recognition of vibrations on his lower limbs, there is no sensory loss. There is no unsteadiness when he stands with his eyes closed.

Wassermann test is strongly positive in blood and cerebrospinal fluid. Cell count in cerebrospinal fluid 22 per cm. Nonne-Apelt test strongly positive.

(11) *A Case of Pain due to a Central Lesion of the Spinal Cord, probably Syringomyelia.*

By Dr. SYMONDS.

The patient, Mr. L. H., aged 45, complains of continuous pain in the left shoulder and arm. He gives the following history:—

Antecedents: He has been employed in the manufacture of water-colours for twenty years. He has had no serious illnesses since childhood, and denies venereal disease. Nineteen years ago he accidentally cut the palmar tendons of his right hand. Thirteen years ago he had an abscess in his left thigh. He has been married twenty-three years, and has three healthy children.

Present illness: About nine years ago he first noticed while at work a feeling of roughness in the skin covering the thumb and first finger of the left hand. "It did not matter what he touched, it felt like a bit of sandpaper." This feeling of roughness later became continuous, being present whether he touched things or not, and gradually spread upwards until about two and a half years ago he experienced the same sensation all the way up his arm, across the left half of the chest above the nipple and over the left shoulder.

Four years ago he burned his left thumb with a poker and a blister was produced without any sensation of heat or pain. This experience has since been repeated on several occasions. In August, 1921, while in hospital, letting his left hand rest upon a hot-water pipe he suffered a severe burn upon the dorsal surface of the forefinger, this being brought to his notice by the appearance of a blister only.

Three years ago he was first aware of some weakness of the left hand

and wrist. This has steadily progressed until he is now unable to use his left hand for handling objects or for doing up the buttons of his clothes. On this account he was obliged to give up work two and a half years ago.

Eighteen months ago he first noticed involuntary jerking movements of the left arm, beginning in the muscles of the shoulder.

Two and a half years ago he began to have pain. At first this was experienced only in the left shoulder, being described as "a tight drawing feeling." This has increased in severity, and as time went on would sometimes shoot down the arm into the limb and first finger. Latterly it has spread to the hand and other fingers.

Two years ago he became aware that when he touched the skin over the lower part of the left face it would cause a shooting pain "like electricity" to travel down the left arm into the finger-tips. He discovered for himself that a light touch or flick of the skin with the finger is most likely to give rise to this painful sensation, or even a touch with the tip of the tongue upon the skin at the angle of the mouth. Continuous pressure is not followed by the same result. He therefore shaves that side of his face with a continued steady pressure of the razor instead of a series of short strokes. He is quite definite in his statement that the skin area from which this reflex pain can be elicited extends exactly to the midline of the face.

Examination of central nervous system: The patient does not give a very clear history, but is quite a fair witness to his sensations.

Special senses: Examination reveals no abnormality.

Cranial nerves: The left pupil is slightly larger than the right, and dilates more rapidly after stimulation with light. Otherwise there is no abnormality in the domain of the cranial nerves, except the sensory changes in the trigeminal field described below (see accompanying charts).

Sensations: Light touch, especially a flick with the finger, applied to the skin of the lower part of the left cheek gives rise to a sensation described by the patient as "an exceedingly painful electric shock," shooting down through neck and shoulder into the left arm and affecting especially the limb and fore-finger. The threshold for the reflex is lowest over the area supplied by the third division of the left trigeminal, and extends exactly as far as the midline in front. The reflex can also be elicited from the lower part of the area supplied by the second division of the trigeminal on this side, but the threshold here is higher.

To cotton wool there is loss of sensation over the left shoulder, and diminution of sensibility not amounting to complete loss over the left arm.

To pin-prick there is complete insensibility of left arm and shoulder, and of the trunk between the second dorsal and ninth dorsal levels. The area of analgesia on the left side extends upwards to include the left side of the back of the head and neck. There is also a subjective diminution to pin-prick over the left side of the face as compared with the right. There is partial analgesia to pin-prick of the right arm affecting mainly the posterior surface and ulnar side. To heat and cold the cutaneous loss follows much the same distribution as that for pin-prick.

Motor system: At rest there are apparent certain involuntary movements mainly confined to the left upper limb. These consist of irregular clonic spasms affecting either single muscles or their fasciculi.

The muscles chiefly affected are the pectoralis major, triceps, biceps, latissimus dorsi, long flexors and extensors of wrists and fingers. On the right side also there are occasional twitchings of the pectoralis major.

There is some wasting of the intrinsic muscles of the left hand and general weakness of the left upper limb affecting mainly the power of hand and finger movements.

Reflexes.—Upper limbs: Tendon jerks on the right are brisk: on the left absent.

Abdominal: All present; the right perhaps a shade brisker than on the left.

Lower limbs: Knee and ankle jerks are brisk, greater on the right than on the left. The plantar reflex on the right is flexor; on the left indefinite. The sphincters and gait are normal.

The Wassermann reaction is negative in blood and cerebrospinal fluid. The cell and protein content of cerebro-spinal fluid are normal. The X-ray plates of the cervical and upper dorsal vertebræ show no abnormality.

This would appear to be a slowly progressive lesion of the central portion of the cord in the upper dorsal and cervical regions, probably a syringomyelia, in which for the past two and a half years continuous pain of a severe character has been the most prominent feature.

I would draw particular attention to the pain of reflex origin produced in the neck, shoulder and arm of the left side by stimulation of the lower trigeminal skin field on the same side of the body.

Notices of Recent Publications.

Anatomie du Système Nerveux. By Cornelis Winkler. Première partie, 1918, pp. 435. Deuxième partie, 1921, pp. 373.

The appearance of the second volume of this anatomy of the nervous system enables us to review the work as a whole; and while we regret that it is not yet complete, and look forward with interest to the appearance of a third volume, there is much in those before us to call for appreciation and comment.

The work is conceived on a large scale, and is intended to give a complete description of the anatomy of the nervous system from the terminal sense organs to the cortex. Nor does it stop there, but it traces the reflex paths along the association fibres of the cerebrum to the efferent tracts of the cord. There for the time being we leave them, but in the promised third volume a fuller description of these efferent paths and their effector organs, as well as of the cerebellum, will, no doubt, be given. The author does not confine himself to human anatomy. For, indeed, an anatomy dealing only with the human nervous system must necessarily be very incomplete, as little is definitely known of many of the tracts and systems in the human nervous system, of which the counterparts have been studied with considerable detail in the lower animals: and it is a mistake too often made in textbooks of anatomy to assume that what is true for the dog and cat must necessarily be true for man. Winkler avoids this error by giving along with descriptions of experimental work the clinico-pathological findings which correlate it with human anatomy. Much of his own work is thus presented to us, and with it we are glad to find numerous references to the work of English neurologists. In many places his observations have led him to conclusions which are not those generally accepted. Thus he considers that all the dorsal roots conducting sensory impulses from the skin enter by Lissauer's zone and end in the substantia gelatinosa of the segment in which they enter the cord. With regard to the descending tracts in the dorsal columns of the cord he considers that the "comma tract" of Schultze certainly, and the long descending tract of Hoche probably, are made up of exogenous fibres, and are, in fact, the prolongations of the descending branches of the dorsal roots. Again, the mesencephalic root of the trigeminal nerve appears to him to be a nerve in the autonomic system of the trigeminal, trochlear and oculomotor nerves. This is in opposition to the teaching of Ariëns Kappers, who considers it to be a part of the sensory root, the ganglion cells of which have not migrated from their original position within the cord. Such instances could be multiplied and

serve to show, what is indeed evident throughout the work, that the author relies chiefly on his own observations in forming conclusions on disputed points of anatomy. But in most instances where his views differ from those of others the reasons for this divergence are fully exposed, and the grounds on which the older views were based fairly stated. The reader is thus presented with data on which to form his own judgment.

The book follows a somewhat novel method of arrangement in that it traces tracts and systems according to their functions rather than their situation. This, though logical, does not always lead to clarity or brevity of exposition. In the first volume the olfactory and optic nerves are first described, along with all their connections with the brain-stem and cortex. Then follows a full description of the peripheral nerves subserving cutaneous sensibility, to which is added a full description of the spinal cord and its reflex paths, and of the connections of the spinal afferent paths with the brain-stem, thalamus and cortex. To this is appended a fairly short description, amounting to only twenty-five pages, of the longer efferent paths of the brain-stem and cord. In this connection he is very guarded as to the importance of the part played in human physiology by the efferent tracts arising in the brain-stem, and says "It is not yet proved, though it seems probable, that there are in the ventral and lateral pyramidal tracts fibres coming from the corpus striatum." This volume closes with two chapters of descriptive anatomy of the brain-stem, and one on the nervous apparatus of taste.

The second volume deals exclusively with the fifth and eighth pair of cranial nerves and their central connections. Of the 260 pages devoted to the eighth nerve, a large number deal with the recent work of Magnus and de Kleyn on the vestibular nerve. Following these authors Winkler provisionally divides the eighth nerve into three main components: (*a*) the cochlear nerve; (*b*) the fibres derived from the *maculæ acusticæ*, of which the part derived from the saccule runs with the cochlear division, and that from the utricle with the vestibular division; and (*c*) the fibres coming from the semicircular canals and ampullæ.

The volumes are well and profusely illustrated. The method of presentation is probably the best for those who, having studied the gross anatomy of the brain and cord, wish to learn their structure in relation to their function. They are emphatically books for the study rather than for the laboratory, and are much more suited for continuous reading than for reference.

J. G. GREENFIELD.

Les Tumeurs du Cerveau. By VIGGO CHRISTIANSEN. Pp. 337. Paris: Masson. 1921.

This book by a Danish author is, fortunately, published in a French translation, and in this form is a very useful addition to neurological literature. Following the modern fashion it takes the form of a series of clinical lectures. It is written in a simple and clear style and leads the reader in ordered sequence from those tumours which present such simple and

unequivocal clinical signs as hemiplegia and aphasia to those in which the signs are more complex and the localization more difficult.

It should therefore be especially valuable to the student of neurology who has not the opportunities given by a large clinic of seeing a great variety of cerebral tumours.

In the earlier chapters the author often seems to take a case of cerebral tumour as a peg on which to hang a general discussion of the function of various parts of the brain cortex, so that the reader is introduced to a wider survey of neurology than if the lectures were strictly confined to the matter in hand. Similarly in dealing with tumours of the base of the brain the author introduces cases of *migraine ophthalmoplégique* and of myasthenia gravis to illustrate the differential diagnosis. Tumours of the hypophysis and the infundibulum, and of the acoustic nerve, are thoroughly discussed from the standpoint of diagnosis. With regard to the latter the author emphasizes the importance of early diagnosis at a stage when the tumour is small enough to be completely removed by operation. He lays stress in the diagnosis of these tumours on a history of vertigo or tinnitus as an early symptom, on the early appearance of papillœdema, and on the presence of an increase of albumin in the cerebrospinal fluid.

In the last chapter but one on "Uncertain Diagnosis" he discusses the reasons why, in many cases, localization of a cerebral tumour may be impossible, and gives details of illustrative cases. The last chapter on "Surgical Treatment" is a lame finish to an excellent book, but like the previous one it shows the author's mental honesty. His own operated cases numbered thirty-three, and of these eleven only had survived up to the time of the lectures. Of these nine were tumours or cysts in the supratentorial region of the cranial cavity, while the other two were tumours of the acoustic nerve partially removed by the translabyrinthine route.

The book is one which will be read with interest by the experienced neurologist, and with advantage by the student of neurology, although it scarcely fills the rôle of a book of reference.

La Dégénérescence Hépato-lenticulaire. By H. C. HALL (of Copenhagen). Pp. 358. Paris: Masson. 1921.

This monograph deals with the diseases in which degeneration of the lenticular nucleus is constantly associated with cirrhosis of the liver. The name hepato-lenticular degeneration thus covers not only progressive lenticular degeneration, but also the pseudo-sclerosis of Westfall and Strümpell, and certain cases of torsion spasm. The author considers that these three diseases are closely related; and, while typical cases are clinically distinct, there are others in which it is not easy to make an exact differential diagnosis.

Progressive lenticular degeneration and pseudo-sclerosis are shown to have certain clinical features in common in addition to those due to disorder of motor function. The most important of these is a peculiar form of corneal

pigmentation due to the deposit of pigment in the outer part of the membrane of Descemet. It is much more commonly found in cases of pseudo-sclerosis than in those of progressive lenticular degeneration, and, so far as is known, does not occur in any other form of disease. Microchemical examination of this pigment proves that it is neither melanin nor lipochrome, nor yet either of the ordinary blood-derived pigments, hæmosiderin and hæmatoidin. On the other hand it corresponds in many particulars to the pigment produced by the activity of the plasmodium of malaria. It responds also to some of the chemical tests for the pigment of argyrism, but Hall considers that it is very improbable that it is derived from silver.

Another clinical feature common to the two diseases is the tendency to alimentary glycosuria, and the constancy, in the cases which have been tested, of alimentary lævulosuria. This tendency is, no doubt, associated with the depraved function of the liver. The nature of the cirrhosis appears to be identical in both diseases, although it has been variously interpreted. Some authors consider it due to faulty development of the hepatic lobules, while others attribute it to some enterogenous toxin. Hall thinks that it is a true cirrhosis, although it differs from the common cirrhosis of Laennec in being much more vascular, and, in consequence, having very little tendency to cause ascites.

He deals at some length with the familial nature of the disease. His own material consists of seven cases, only one of which was followed to autopsy. These seven cases were derived from three families, two from one and four from another. The latter family is of great interest inasmuch as the cases occurred in two generations, with a common male ancestor—two sons of a daughter by a first marriage and a son and daughter by a second marriage being affected. From this he argues that the disease is probably due to the fusion of two hereditary trends, one of which occurs very rarely and shows itself as a dominant, whereas the other occurs fairly commonly, and may be either dominant or recessive. In the instance quoted above the first hereditary trend would be present in the father, and the second in his second wife and the husband of the daughter of the first marriage.

The author deals fully with the pathology of the cerebral degeneration, not only detailing his own findings but giving a complete résumé of those of other workers. From this it appears that the degeneration is by no means confined to the lenticular nucleus, although it has its greatest intensity there. In pseudo-sclerosis particularly changes, especially of the nature of glial overgrowth with the formation of abnormal types of glial cell, were found in parts as distant as the cortex and the pons.

He deals somewhat scantily with the theories as to the physiological basis of the motor symptoms. In this he is probably wise, as our knowledge of the physiology of the corpus striatum still rests on somewhat insecure foundations.

The book forms a valuable addition to neurological literature in that it presents in readable form a full summary of work which up to now has been scattered in many lengthy papers in various languages. And the author's wise judgment and critical acumen add greatly to its value.

The Psycho-analytic Study of the Family. By J. C. FLÜGEL, B.A. (The International Psycho-analytical Library No. 3). Pp. 259. London, Vienna, New York: International Psycho-analytical Press. 1921.

"A person whose instincts and impulses are co-ordinated sufficiently to maintain, as regards all the leading aspects of life, a relatively harmonious functioning of the whole personality can preserve mental health in circumstances under which a less integrated mind would fail." One of the most potent influences to which a human being is exposed from the cradle to the grave is family life. The object of this book is to study this factor in its various forms as it appears in abnormal psychology. Primitive emotions, conflict, the growth of individual personality, love and hate, are all examined from this point of view. The effect of early family influences on social development, religion, the attitude of parents to children and the various forms which may be assumed by sexual desire are described according to the teaching of Freud and his followers; the views of Jung receive due attention, though they are not accepted by the author. The book contains no original investigations, but is a useful compendium of the theories of the psycho-analytical school on the diverse manifestations produced by family life in the psychology of the individual.

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